Journal of Current Hematology & Oncology Research

Case Report

DOI: 10.51271/JCHOR-0016

# A rare case of acute basophilic leukemia

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**Cite this article:** Çelik S, Keklik M, Kontaş O, Koker MY, Kaynar L. A rare case of acute basophilic leukemia. *J Curr Hematol Oncol Res.* 2023;1(3):73-75.

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**Received**: 23/06/2023 ◆ **Accepted**: 11/07/2023 ◆ **Published**: 29/08/2023

## ABSTRACT

Acute basophilic leukemia is an extremely rare form of acute myeloid leukemia and diagnosis is based on morphological, cytochemical and ultrastructural analyses of basophils and blastic cells. It is usually characterized by a very rapid clinical course, symptoms of hyperhistaminemia and resistance to therapy. A 73 year-old male presented with itching, thrombocytopenia and leukocytosis. There was no hepatosplenomegaly. In the flow cytometry evaluation of peripheral blood, CD123 positive myeloid blasts were detected at the rate of 60%. Also, CD13 and CD33 were positive, while CD34, CD117, HLA DR and myeloperoxidase (MPO) were negative. These findings were considered as consistent with ABL. Following, the diagnosis of ABL was confirmed by bone marrow evaluation including cytomorphological and immunohistochemical studies. On the other hand, cytogenetical analyses of bone marrow showed the presence of the Philadelphia chromosome. The patient received one cycle of a chemotherapy regimen including cytarabine and idarubicin with imatinib support, but he died on the 10th hospital day. Considering that ABL behaves in a very malignant fashion, and a more aggressive treatment approach is necessary for patients with this specific subtype of AML.

**Keywords**: Acute basophilic leukemia, CD123 antigen, flow cytometry

## INTRODUCTION

Acute basophilic leukaemia (ABL) is an uncommon form of acute myeloid leukemia (AML) accounting for 4-5% of all cases.[1] Consistent diagnostic criteria for ABL still remain the topic of discussion. For diagnosis, it is necessary to exclude other hematological malignancies associated with basophilia such as chronic myeloid leukemia (CML), acute promyelocytic leukemia (APL) with basophilic differentiation, and AML/myelodysplastic syndrome (MDS). On the other hand, most cases of ABL have been reported to be a consequence of the evolution from other leukemias. In most cases, the prognosis is poor. Here, we report a rare case of ABL with a very agressive clinical course.

## CASE

A 73-year-old male presented to hematology department with itching lasting for 3 months. He had no past any medical history, family history and history of tobacco, alcohol, illegal drug use, and travel. Also his past medical history was negative for atopic diseases. His vital signs were blood pressure 110/70 mmHg, pulse 88/min, respiratory rate 23/min, and body temperature 36.7 °C on admission. There was no hepatosplenomegaly or lymphadenopathy. Laboratory studies revealed the following: hemoglobin of 13.8 g/dl, white blood cells of $63x10^9$/L, and platelets of $84x10^9$/L. Serum biochemistry and coagulation were unremarkable.

There were large population of basophils on the peripheral blood smear. Viral, bacteriological and autoimmune evaluations were normal. Also, cytomegalovirus, rubella virus, herpes simplex virus, varicella zoster virus, Epstein–Barr virus, hepatitis markers and brucellosis tests were found as negative. In addition, analysis to detect a parasitic infection or an atopic cause were negative.

Spleen size was normal in the abdominal ultrasonography. Following, flow cytometry (FC) evaluation was performed by using FACS Calibur flow cytometer (Becton–Dickinson, Erembodegem, Belgium). In the FC evaluation of peripheral blood, CD123 positive myeloid blast cell group was detected at the rate of 60% of the total localized at CD 45 dim (Figure-1).

CD4: CD8 ratio was 65:33. Also, CD13 and CD33 were positive, while CD34, CD117, HLA DR and myeloperoxidase (MPO) were negative. These results were considered as consistent with ABL. Following, bone marrow biopsy was performed, and the aspirate revealed high cellular marrow and most of the cells were CD13 and CD33 positive (**Figure 2**).

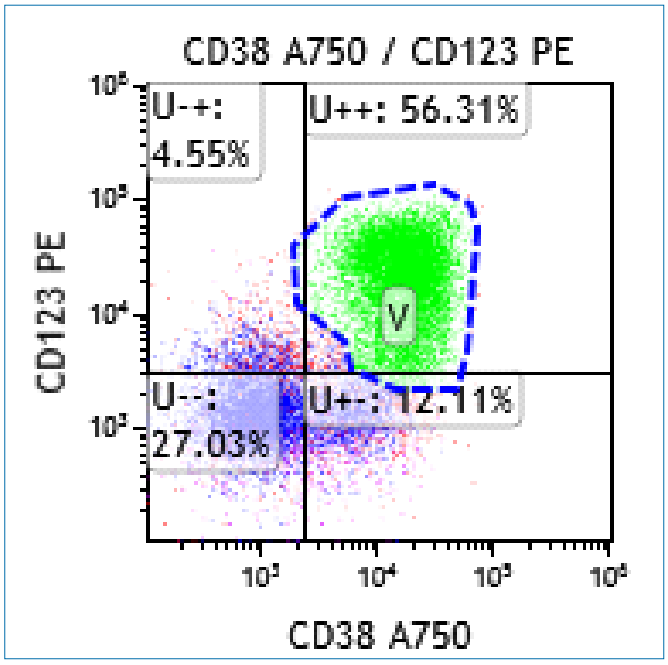


Figure 1. Shown is the 60% of CD-123 positivity in flow cytometry.

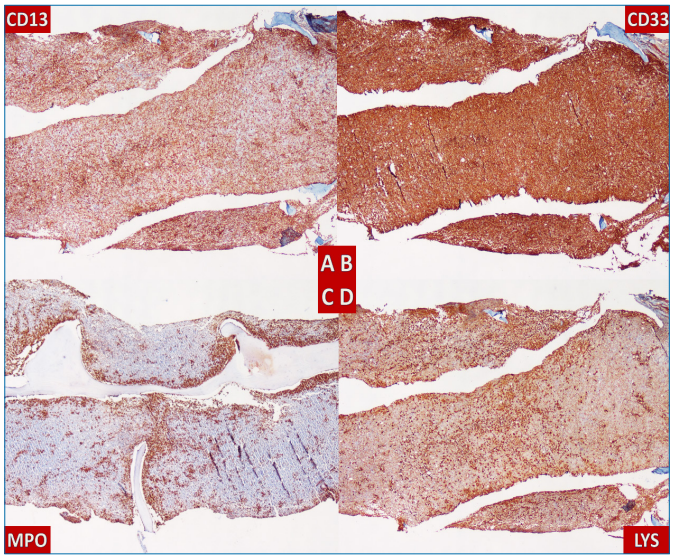


**Figure 2.** Immunohistochemical results. Most of the bone marrow cells are CD13 (A) and CD33 (B) positive. MPO is positive(C) in only residual normal granulocytes but lysosyme is positive (D) in both in normal granulocytes and some neoplastic cells (all sections are x40).

MPO was positive in only residual normal granulocytes but lysosyme was positive in both in normal granulocytes and some neoplastic cells. Also, the blasts were negative for CD34, CD117, CD10, TdT and glycoforin. In the histopathologic sections of biopsy, mature erithroid and granulocytic cell ratio was diminished and some cells with abundant cytoplasm, indented ovoid nuclei were raised. There was a mild reticulin fibrosis and there was no siderophage in bone marrow. With these findings, bone marrow biopsy was reported as ABL.

Cytogenetic studies performed on bone marrow aspirate using standart culture methods and banding technique revealed t(9;22) and BCR/ABL1 fusion gene was detected by FISH analysis.

The patient was treated with a standart 7+3 regimen including cytarabine and idarubicin with imatinib support. Also, he received histamine receptor (HR) antagonists (HR1 and HR2 blocker) for itching. However, he died on the 10th hospital day due to aggressive progression of ABL.

## DISCUSSION

Acute basophilic leukemia (ABL) has been known for a century. However, consistent diagnostic criteria are lacking. Therefore, most cases previously reported as ABL represented a variant of CML or developed secondary to MDS. Following, de novo ABL cases have been reported in recent years.

Later, WHO classifications, ABL has been integrated as a ABL is an aggressive and extremely rare type of leukemia and is now classified as a distinct entity in the WHO classification. entity and defined as an AML in which primary differentiation is towards the basophil lineage.[2] However, no generally accepted criteria for ABL have been presented to date. To address this issue, in 2017, Valent et al.[3] proposed a diagnostic algorithm of basophilic leukemias. According to their recommend, for the diagnosis of basophilic leukemia, the percentage of basophils must be ≥40 of total leukocytes. In our case, this ratio was 60%. Also, they suggested the CD123 positivity for the diagnosis. In our case, CD123 (+) blastic cells group ratio was 60% in the FC analysis. In addition, they suggested to considered other causes of basophilia such as chronic infections, autoimmune processes and atopic disorders. In our case, investigations to detect these possible causes were negative. Also, they proposed the bone marrow examination for differential diagnosis of ABL. In our case, bone marrow biopsy resulted as ABL.

On the other hand, the Philadelphia chromosome had been described in some cases of MDS.[4,5] In our case, there were no any findings of cytomorphological evaluations compatible with MDS.

Actually, CML is the first diagnosis proposed in our patient due to positive Philadelphia chromosome, however, there was no splenomegaly, and the patient did not have positive history to CML, on the other hand, the rapidly progression, positive myeloid markers (CD13 and CD33), and positive CD123, make the diagnosis of ABL. Already, the diagnosis was confirmed by detailed bone marrow evaluation including cytomorphological and immunohistochemical studies. Even if our patient was diagnosed with CML, he still had ABL diagnosis according to current criterias. However, in this case it is called secondary ABL. In any case, whether it was primary or secondary, he had a diagnosis of ABL.

According to the literature, the survival time in ABL patients ranges from 2 to 16 months.[6] Therefore, these patients are often regarded as candidates for allogeneic stem cell transplantation (ASCT). However, not all cases are eligible for ASCT. Also, there are no prospective trials comparing treatment options for ABL. Several authors have reported results of various induction therapies for ABL. These have included treatment used for AML and acute lymphoblastic leukemia. However, the results have been disappointing.[7] Similarly, our case received one cycle of induction chemotherapy including cytarabine and idarubicin, but he died on the 10$^{th}$ hospital day due to aggressive progression of ABL.

## CONCLUSION

Due to these results found in our case, ABL behaves in a very malignant fashion, and more aggressive treatment approach is necessary for this patients.

### Learning Points

ABL is an aggressive and extremely rare type of leukemia and is now classified as a distinct entity in the WHO classification.

It is important to differentiate ABL from CML, as in our case, the percentage of basophils above 40, CD 123 positivity and the absence of hepatosplenomegaly are important in the diagnosis of ABL.

There is still no clear algorithm in ABL treatment approach, so further studies are needed in this area.

## ETHICAL DECLARATIONS

**Informed Consent:** All patients signed the free and informed consent form.

**Referee Evaluation Process:** Externally peer-reviewed.

**Conflict of Interest Statement:** The authors have no conflicts of interest to declare.

**Financial Disclosure:** The authors declared that this study has received no financial support.

**Author Contributions:** All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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