

Case Report

Atypical Chronic Myeloid Leukemia - A Rare Clinical EntityPatil Nanda J¹, Dhawan Shrutika D², Borade Pramod D², Gosavi Anup N²¹Professor, ²Tutor,

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ABSTRACT

Atypical chronic myeloid leukemia (aCML) is a rare myeloproliferative neoplasm with distinct haematological and cytogenetic characteristics than classical chronic myeloid leukemia cases. The disease has poor clinical outcome. We present this case to highlight its characteristic features.

Key words: CML, atypical, Philadelphia negative.

INTRODUCTION

Atypical chronic myeloid leukemia (aCML) is a rare myelodysplastic / myeloproliferative neoplasm with an estimated relative incidence of 1 to 2 cases for 100 patients with BCR-ABL-1 positive chronic myeloid leukemia. ^(1,2) It has distinct haematological and cytogenetic characteristic which differ from classical chronic myeloid leukemia (CML). These cases have poor clinical outcome. ^(1,3,4)

CASE HISTORY

A 72 years male patient presented with generalized weakness and abdominal pain. On clinical examination, moderate splenomegaly was noted. Haematological investigations revealed anemia (Haemoglobin-8.6gm/dl); leucocytosis (TLC-21,700/mm³) with differential count - 86% myeloid series cells; 2% Blast cells; 2% basophils and 10% lymphocytes. Myeloid series showed shift to left with

dysplastic features that included hypogranular and hypolobated neutrophils having irregular nuclear chromatin (Fig 1 & 2). Red cell count was decreased (3.40millions/mm³). Platelet count was within normal limits.

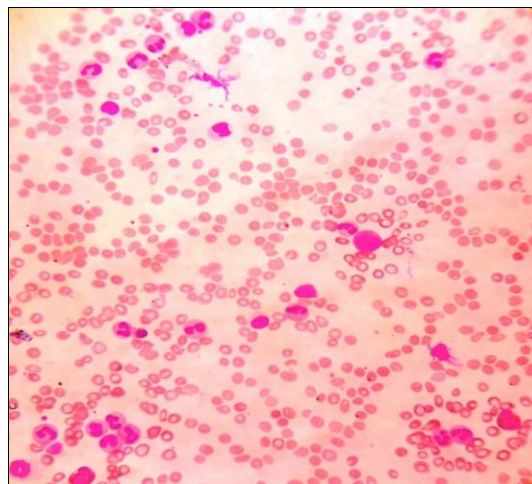


Fig 1: Photomicrograph of peripheral blood smear showing leucocytosis with increased immature and dysplastic granulocytes (100x, Giemsa).

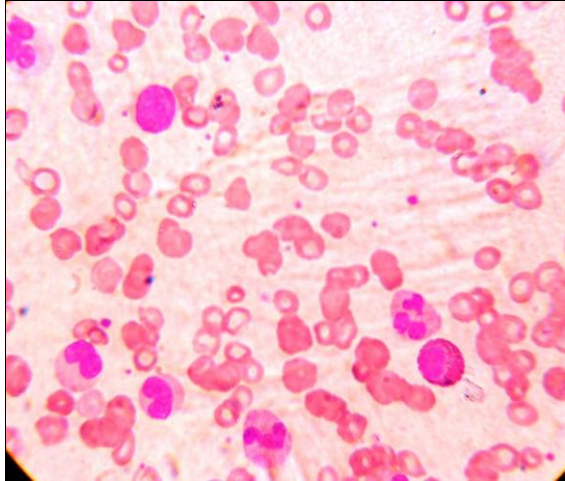


Fig 2: Photomicrograph of peripheral blood smear showing granulocytic dysplasia i.e. hypolobated and hypogranular neutrophils, a single blast and hypolobated eosinophil (100x , Giemsa).

Bone marrow aspiration study revealed hypercellular marrow with predominance of myeloid cells showing dysplastic features. Megakaryocytic series also showed dysplastic features.

Leucocyte Alkaline Phosphatase (LAP) score was 280 (Normal-40 to 140). The cytogenetic study was negative for Philadelphia chromosome as well as BCR-ABL rearrangement.

DISCUSSION

aCML is a rare entity having distinct haematological and cytogenetic features than classical CML. These cases present with splenomegaly, neutrophilic leucocytosis showing minimal left shift. Granulocytic series revealed dysplastic features like hypogranular and hypolobated neutrophils; nuclear chromatin clumping and pseudo Pelger-Huet neutrophils. ⁽¹⁾ The total leucocyte count is often more than $13,000/\text{mm}^3$ with immature granulocytes more than 10% of leucocytes. The percentage of blast is less than 20% in blood smear and bone marrow. Differential basophil count is less than 2% and monocyte count is less than 10% in peripheral blood smear. ⁽⁵⁾ Majority of the patients showed

red cell hypoplasia. ⁽⁶⁾ Our case presented with splenomegaly and same haematological observations.

Leucocyte Alkaline Phosphatase (LAP) level in these patients may be low, normal or increased; hence lack the diagnostic utility. ^(3,4) In our case LAP score was increased upto 280. aCML cases reveal hypercellular bone marrow with myeloid hyperplasia with prominent granulocytic dysplasia. Multilineage dysplasia may be present. ⁽⁶⁾ Bone marrow features in our case were increased cellularity, myeloid hyperplasia with dysplasia in granulocytic and megakaryocytic series.

The cases of aCML have cytogenetic characteristic which differ from classical CML cases. The Philadelphia Chromosome which is considered as hallmark of CML is absent in aCML cases. Also BCR-ABL rearrangement which is seen in classical CML is absent in aCML. ⁽⁷⁾ The cytogenetic studies in our case were Philadelphia Chromosome negative with no rearrangement of BCR-ABL gene.

The differential diagnosis for aCML includes leukaemoid reaction and Chronic myelomonocytic leukemia. ⁽⁸⁾ Cases of aCML have poor overall median survival which ranges from 14-30 months; few cases transform to acute leukaemia. Shorter survival is associated with old age (> 65 years), female gender, leucocyte count more than $50 \times 10^9/\text{litre}$ and presence of immature circulating precursors. ⁽⁵⁾ The therapy for aCML consists of Hydroxyurea; other chemotherapy well as Interferon- α which results in improved blood counts. ⁽⁵⁾ Our case is stable with treatment by Hydroxyurea and is on regular follow up since one year.

CONCLUSION

Atypical Chronic Myeloid Leukemia is a rare form of leukemia which shows

myelodysplastic and myeloproliferative features simultaneously.

Haematological work-up with cytogenetic studies is essential for making this diagnosis.

Patient presenting with splenomegaly, leucocytosis and dysplastic features in granulocytic series without monocytosis should be considered for diagnosis of atypical chronic myeloid leukemia.

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