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*Case Report*

# A Rare Case Report on T- Cell Lymphoid Blast Crisis of CML with an Unusual Presentation

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## ABSTRACT

T-cell lymphoid blast crisis is a very rare presentation in chronic myelogenous leukaemia (CML). Considering its rare entity, herein we report a case of T-cell lymphoid blast crisis of CMLin a 27 year old female who presented with anaemia, leucocytosis and thrombocytosis. The patient has no previous history of CML and the onset of CML blast crisis was sudden.
The unusual presentation is that the patient had a refractory thrombocytosis for which splenectomy was done 8 months back and at that time no other hematologic abnormalities were detected. The patient succumbed to death within few months of diagnosis.

***Key-words***: CML, T-cell, lymphoid blast crisis

## INTRODUCTION

Chronic myelogenous leukaemia is a myeloproliferative neoplasm that is associated with BCR-ABL1 fusion gene located in the Philadelphia chromosome. [1] CML almost always demonstrates the p210 fusion protein. The typical clinical course is an indolent chronic phase followed by blast crisis or progression to an accelerated phase with subsequent blast crisis. Although most blast crisis are myeloid blast crisis, one third are lymphoid of which 90% are of B-cell type and rarely T-cell blast crisis have been reported.

## CASE REPORT

A 27 years old female patient presented with haemoglobin 6gm% and total white blood cell count 30,000/cu.mm and total platelet count 10.2lakhs/cu.mm. She had a previous history of splenectomy 8 months back due to refractory thrombocytosis. Hemolytic anemia by HPLC was also ruled out then.

A bone marrow study showed cellular aspirate with 30% lymphoid blast cells and myelocytes and metamyelocytes are also present. Erythropoiesis is depressed with M:E ratio 18:1. Megakaryocytes are present, normal in numbers, mostly mature and packed with platelets. Marrow picture was suggestive of acute leukemia.

Flow cytometry demonstrated 45% blast cells which express CD7(dim), CD13 (partial/dim), CD33 (dim), CD34 (heterogenous) and cytoplasmic CD3. All other markers tested including HLA-DR and cytoplasmic MPO are negative in blasts. So flow cytometry was suggestive of a diagnosis of early T-precursor lymphoblastic leukemia.

Molecular analysis showed positive BCR-ABL transcript of p210 fusion gene protein by RT-PCR (BCR-ABL1/ABL1% Ratio: 33.5194).
A diagnosis of early T-precursor lymphoblastic leukemia as blast crisis in CML was made based of above flow

cytometry and molecular analysis. The patient succumbed to death after few months of diagnosis.

Leukaemoid reactions and other myeloproliferative neoplasms are ruled out.

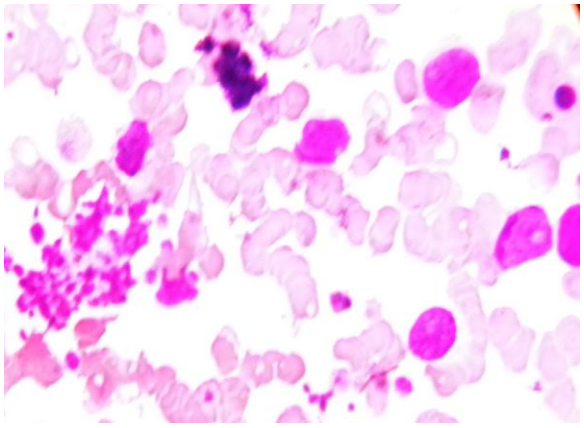

**Figure 1 Bone marrow showing lymphoid blast cells and platelet clumps**

## DISCUSSION

Chronic myelogenous leukaemia (CML) is a myeloproliferative neoplasm that originates in an abnormal pluripotent bone marrow stem cell and is consistently associated with the BCR-ABL1 fusion gene located in the Philadelphia chromosome. [1]

CML has a worldwide annual incidence of 1-2 cases per 100,000 population with 5$^{th}$ and 6$^{th}$ decade as mean age of diagnosis and slight male predominance. [2]

20-40% of patients are asymptomatic and diagnosed on routine blood examination done for pre-operative or other purposes. [2,3] Common clinical presentations include fatigue, weight loss, night sweats, splenomegaly and anemia. [3,4] Majority of Indian patients are symptomatic and mostly present with dull aching pain in the left hypochondriac region secondary to splenomegaly. [5,6]

In approximately 70% of cases, the blast lineage in blast crisis is myeloid and in 20-30% lymphoblasts. [7-12]

In myeloid BP, the blasts may or may not express MPO but express antigens associated with granulocytic, monocytic, megakaryoblastic, erythroid and at times one or lymphoid antigens. [7-9,11] In lymphoid BP, 90% are of B-cell origin but rare T-cell origin is reported. [8,9,11] Mixed phenotypic acute leukemias are also reported in BP. [7-9,11]

CML almost always demonstrates the p210 fusion protein. p230 fusion protein is typical of the rare neutrophilic form of CML and p190 in t(9;22) of acute leukemias. Very rarely p190 BCR-ABL positive CML is described.

Blast crisis in CML depicts a clonal evolution of abnormal cells that have gathered during the chronic phase. [13] Most CML cases are diagnosed during the chronic phase whereas some CML patients present during blast crisis. Lymphoid blast crisis constitutes around 25% of blast transformation in CML. [14,15] Most lymphoid blast crises are of B cell type with very few reported T cell phenotypes. [15] Sudden onset blast crisis is, however, more common with lymphoid than myeloid blast transformation. [2]

The patients are usually adults with a male predominance. Most cases have a history of CML or concurrent CML in bone marrow. In all reported cases of T- cell lymphoid CML blast crisis, BCR-ABL1 gene fusion occurs at the BCR major breakpoint with a protein product of p210. Most patients show a poor prognosis.

The most important differential diagnosis is de novo T-ALL in adults which can be ruled out on basis of occurrence of p190 BCR-ABL1 gene product in ALL.

The case reported here has no previous history of CML and the onset of CML blast crisis was sudden. The patient succumbed to death within few months of diagnosis which shows poor prognosis in sudden onset lymphoid blast crisis of CML. The unique feature is that patient had a refractory thrombocytosis for which splenectomy was done 8months back and at that time no other hematologic abnormalities detected in the patient.

In summary, T-cell lymphoid blast crisis of CML is rare and few cases have been reported. But such sudden presentation and splenectomy history is not reported before. Splenectomy can be hypothesized to

worsen the course of CML and lead to poor survival for the patient.

Because of its peculiar presentation and rarity, the case has been reported.

## CONCLUSION

Only a handful of cases have been reported so far in literature. This is being reported because of the rarity of the case.

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