Case Report: International Journal of Clinical Research and Reports

T Cell Blast Crisis transformation of CML: A Case Report

Temesgen Assefa Ayele[1*], Natnael Fekadu Tekle[2]

[1] Dr. Temesgen Assefa Ayele, Internist, Clinical Hematologist, Hematology Unit, Department of Internal Medicine, Saint Peter specialized Hospital
[2] Dr. Natnael Fekadu Tekle, Internal medicine resident, Adissababa university

***Corresponding Author:** Temesgen Assefa Ayele, Internist, Clinical Hematologist, Hematology Unit, Department of Internal Medicine, Saint Peter specialized Hospital, Adissababa university.
Received: 14 September 2023 | **Accepted:** 25 September 2023 | **Published:** 06 October 2023
Citation: Temesgen Assefa Ayele, Natnael Fekadu Tekle (2023), T Cell Blast Crisis transformation of CML: A Case Report Author Information, *J. International Journal of Clinical Research and Reports*. 2(5); DOI:10.31579/2835-785X/020

Abstract:
Chronic myeloid leukemia (CML) a myeloproliferative neoplasm characterized by the dysregulated production and uncontrolled proliferation of mature and maturing granulocytes with fairly normal differentiation. Clinically it presents in 3 phases: chronic, accelerated & Blast crisis phase. About 5% of cases are diagnosed in blast phase which in 70 % of cases Myeloid & the remaining 30 % are lymphoid blast crisis.
We present a 54 years old male patient with a Chronic phase CML taking Imatinib presented with cervical lymphadenopathy & diagnosed to have a T –cell CML blast crisis.
Key words: CML; blast crisis; T cell

Introduction:

Chronic myeloid leukemia (CML) is a myeloproliferative disorder associated with the Philadelphia chromosome t (9;22) (q34; q11) and the BCR-ABL fusion gene.
Clinically CML manifests in three phases these include the chronic phase which is the most indolent form of the disease & accounts for about 85 -90 % of cases of the disease, the most aggressive forms of the disease are the accelerated & blast crisis accounts for 10% & 5% of CML cases respectively.[1].
CML usually progress from the most stable chronic phase of the disease to the most advanced phase blast crisis with in a period of 4-5 years without treatment.
The advent of Tyrosine kinase inhibitors significantly delayed & /or avoided transformation of the disease to advanced forms in the majority of Chronic phase CML cases. [2].
Certain patients, however, experience resistance to these medications; this occurs through several mechanisms including the accumulation of additional cytogenetic abnormalities, which can confer a survival advantage to the treated myeloid cells & leads to disease transformation to accelerated & blast crisis. The most common cytogenetic abnormalities include an additional Ph chromosome, trisomy 8 and is chromosome 17q. [3].
WHO defines CML blast crisis by the presence of one or more of the following findings: ≥20 percent peripheral blood or bone marrow blasts, large foci or clusters of blasts on the bone marrow biopsy, presence of extramedullary blastic infiltrates.
In the majority of cases, blasts are of myeloid origin accounting for about 70% of cases & Lymphoblasts are seen in 30% of CML blast crisis phases. [4].
Lymphoid blast crisis resembles acute lymphoid leukemia with Philadelphia chromosome positivity. Most reports of CML blast crisis with lymphoid transformation reports a B cell transformation but reports on T cell lymphoid blast crisis are sparse.[5].
Here we report a 54 years old male patient with Chronic phase CML who was transformed to a T cell Blast crisis evidenced by morphologic & Immunologic studies.

Case Presentation:

A 54 years old male patient who is a known chronic phase CML patient on imatinib 400 mg once per day for past 3 years who has regular follow-up & was adherent to his medications & was in a complete hematologic remission since 3months back.

Currently presented with a 3 months history of progressively enlarging bilateral cervical swelling without axillary inguinal or abdominal swelling

He also has symptoms of easy fatigability; tinnitus & vertigo associated with the cervical swelling but has no history of fever or history of bleeding from any site.

On examination he has multiple firms, nontender cervical Lymphadenopathies with the largest measuring 3X 3 cm without other abnormal physical examination findings.

His laboratory parameters revealed WBC – 4.2 X 10^3, ANC -2.6 X 10^3, Hgb- 13.7, platelet -184 X 10^3, LDH – 219, Cr-1.0, Total bilirubin – 0.7, Direct bilirubin -0.1

AST - 42, ALT – 41, AlP - 125

LN Biopsy – Monotonous predominantly medium to occasionally large lymphoid cells with coarse hyper chromatic pattern, inconspicuous nucleus & frequent mitosis with starry sky pattern

LN biopsy with IHC revealed T cell Lymphoblastic Lymphoma with tumor cells that are strongly positive for CD3, Tdt, CD7, CD45 & focally positive for CD 5 & negative for CD34, CD117, Ki67 -80%.

Bone marrow Biopsy with IHC study revealed T lymphoblastic lymphoma with sheets of atypical medium sized lymphoid cells which are diffusely and strongly positive for CD3, TdT, and CD45, focally positive for CD5 & negative for CD20, CD34, and CD117.

BCR –ABL was 95 % determined by FISH, other cytogenetic abnormalities were not detected.

Discussion:

It is well established that ~30% of blast crises in CML are of the lymphoid rather than the myeloid phenotype & among the le lymphoid phenotypes the B lymphocytes are the usual reported phenotype of CML lymphoid blast crisis although the exact proportion of T lymphoid blast crisis is not known.[5].

Exact identification of the blast phenotype has its own therapeutic implications, since the treatment protocol of lymphoid blast crisis is different to that of the myeloid type.

In myeloid blast crisis the preferred initial treatment is the use of a TKI followed by an allogeneic HCT for eligible patients since it is typically refractory to chemotherapy, the main plan in treatment of Myeloid blast crisis should be to return patients to an earlier phase of the disease & subsequently undergoing allogenic stem cell transplantation.[6].

CML lymphoid blast crisis is usually treated similarly with Philadelphia positive acute lymphoblastic Leukemia (ALL) with tyrosine kinase inhibitors in combination with chemotherapy or steroids. [6].

In our particular case the patient transformed in to blast crisis CML while he was on imatinib treatment, so he was started on Dasatinib 140mg daily in combination with Dexamethasone & vincristine since the patient has a poor performance status & was considered unfit for intensive ALL chemotherapeutic regimen. With this treatment the patient achieved improvement in clinical symptoms with disappearance of the cervical lymphadenopathy but didn't achieve in disappearance of the peripheral & bone marrow lymphoid blast after 3 months of treatment.

Conclusion:

Although lymphoid blast transformation of CML is rare we should have high index of clinical suspicion when chronic phase CML patients develop lymphadenopathy.

T – Cell Blast crisis is extremely rare that the exact incidence is not clearly known, we present this very rare clinical condition to show the importance of doing immunohistochemistry & genetic studies to know exactly the type of CML blast crisis.

Disclosure:

- Verbal Informed consent was obtained from the patient

- The patient is fully volunteer for the reporting of the case and to continue the care.

Affiliation:
Saint Peter specialized Hospital

Conflict of Interest
None

References

1. Nada Naiyer, et al. (2022). A rare case of B-lymphoid blast phase of chronic myeloid leukemia: Diagnostic. challenges. *Leukemia Research Reports*,17;100327
2. Bruno Calabretta et al. (2004). The biology of CML blast crisis. *Blood*,103(11): 4010–4022.
3. Hamoud Al-Khallaf, et al. (2018). Precursor B cell lymphoid blast crisis of chronic myeloid leukemia with novel chromosomal abnormalities: A case report. *Oncol Lett.*16(5): 6691–6696.
4. Binoy Yohannan, et al. (2022). B-Lymphoid Blast Phase–Chronic Myeloid Leukemia: Current Therapeutics. *Int J Mol Sci*.23(19): 11836.
5. Vindhyaprabha Hegde, et al. (2020). Flow Cytometry in Chronic Myelogenous Leukemia Blast Crisis. Iranian Journal of Blood & Cancer. *IJBC*,12(3): 80-83
6. Mhairi Copland, et al. (2022). Treatment of blast phase chronic myeloid leukaemia: A rare and challenging entity. *Br J Haematol.* 199(5): 665–678.