MAR Pathology & Clinical Research (2023) 2:3

Case Report

Adult Type Chronic Myeloid Leukemia in a 5 Year Old Child – A Rare Case

Maruti a Dhakane [1], Ankita Majumder *[2], Vandana Gite [3]

*Correspondence to: Ankita Majumder.

Copyright

Received: 26 June 2023

Published: 01 July 2023

Abstract

Chronic myeloid leukemia, a hematopoetic clonal stem cell disorder, is a type of chronic myeloproliferative disorder which is quite rare in paediatric population, amounting upto 2%-5% of all leukemia in this age group. Here, we report a case of CML in a five year old girl child with a clinical presentation of fever and abdominal distension. The diagnosis was made based on bone marrow aspiration findings and molecular study.

Key words: *Chronic myeloid leukemia, paediatric.*

Introduction

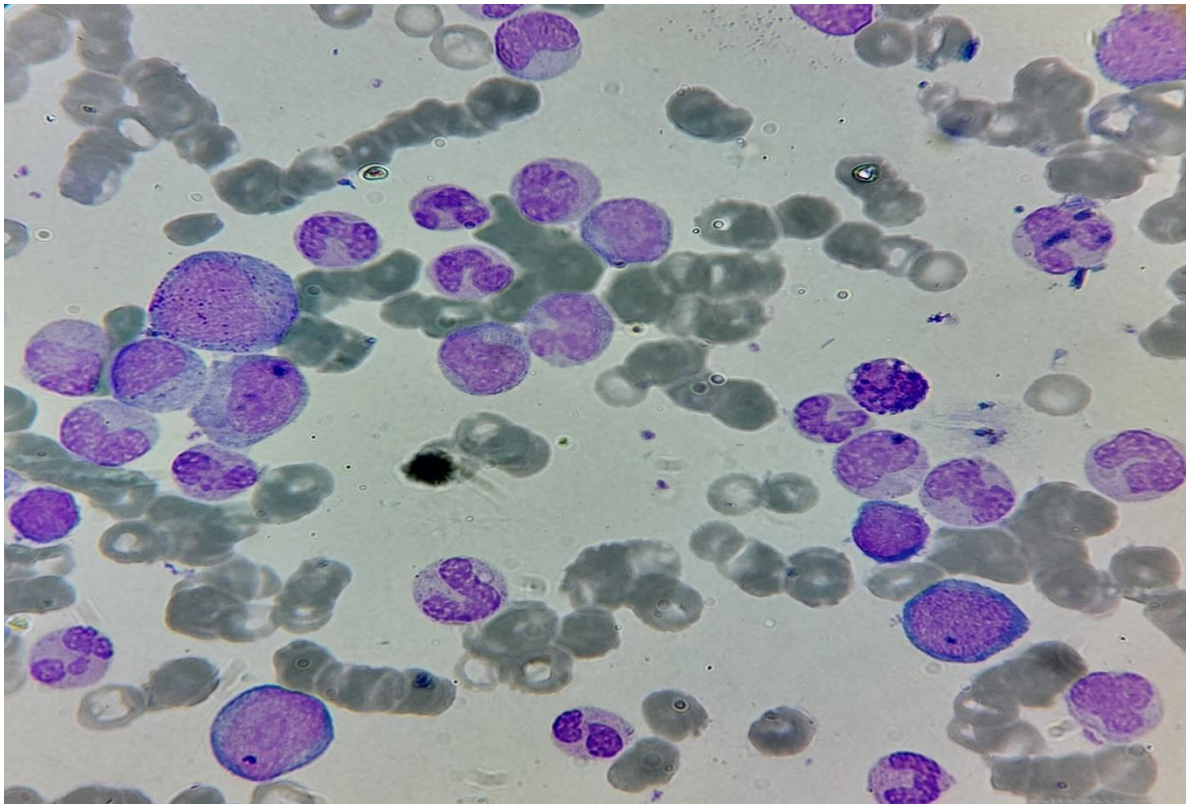
Chronic myeloid leukemia (CML) is a myeloproliferative disorder characterized by the presence of the BCR/ ABL1 fusion transcript encoded by the Philadelphia (Ph) chromosome, a result of a reciprocal translocation between chromosomes 9 and 22 (1). The median age of diagnosis of CML is 60 to 70 years and is rare among paediatric population. CML constitutes 2% of all leukemias in children younger than 15 years with an annual incidence of 1 case per million (2). Recent data highlight the distinct biological differences between adult and pediatric CML. Pediatric CML patients typically have higher mean WBC counts, more pronounced splenomegaly, and pursue a more aggressive clinical course when compared to adult CML patients (3). Here, we report a case of CML in a five year old girl because of its uncommon incidence in this age group.

Case Presentation

Five year old female child came to our pediatric OPD with chief complaints of fever and abdominal distension since 1 month. The child had an uneventful past and was fully immunized according to her age. On examination,the averagely built child had hard and distended abdomen. Liver was palpable 6 cms and huge splenomegaly was noted of 20 cms. In systemic examination no significant abnormality was detected. The patient was admitted in pediatric ward with differential diagnosis of malaria or storage disorder. Routine hemogram was carried out, which revealed Hb – 8.3 gm/dl with markedly elevated Total leukocyte count

of 2, 65,700/Cumm. A peripheral differential count revealed shift of myeloid series up to myeloblasts. Basophilia was noted. (Polymorphs + Band forms = 52%, Metamyelocytes = 10%, Myelocyte = 21%, Promyelocytes = 02%, Blasts = 03%, Lymphocytes = 02%, Eosinophils = 06%, Monocytes = 01%, Basophils = 03%) (Fig. 1, 2). Platelet count was normal with 2.85 lac/ Cumm . On peripheral smear examination, the diagnosis of - Chronic Myeloproliferative disorder suggestive of CML (adult) type was given.

Cytogenetics was advised and the result came positive for BCR-ABL gene fusion, BCR-ABL1 Ratio % - 11.92%, BCR-ABL1 copy number 21,000; BCR-ABL!/ABL ratio 16.79%. It supported and confirmed the diagnosis of chronic myeloid leukemia of adult type and the patient subsequently was started on chemotherapy.



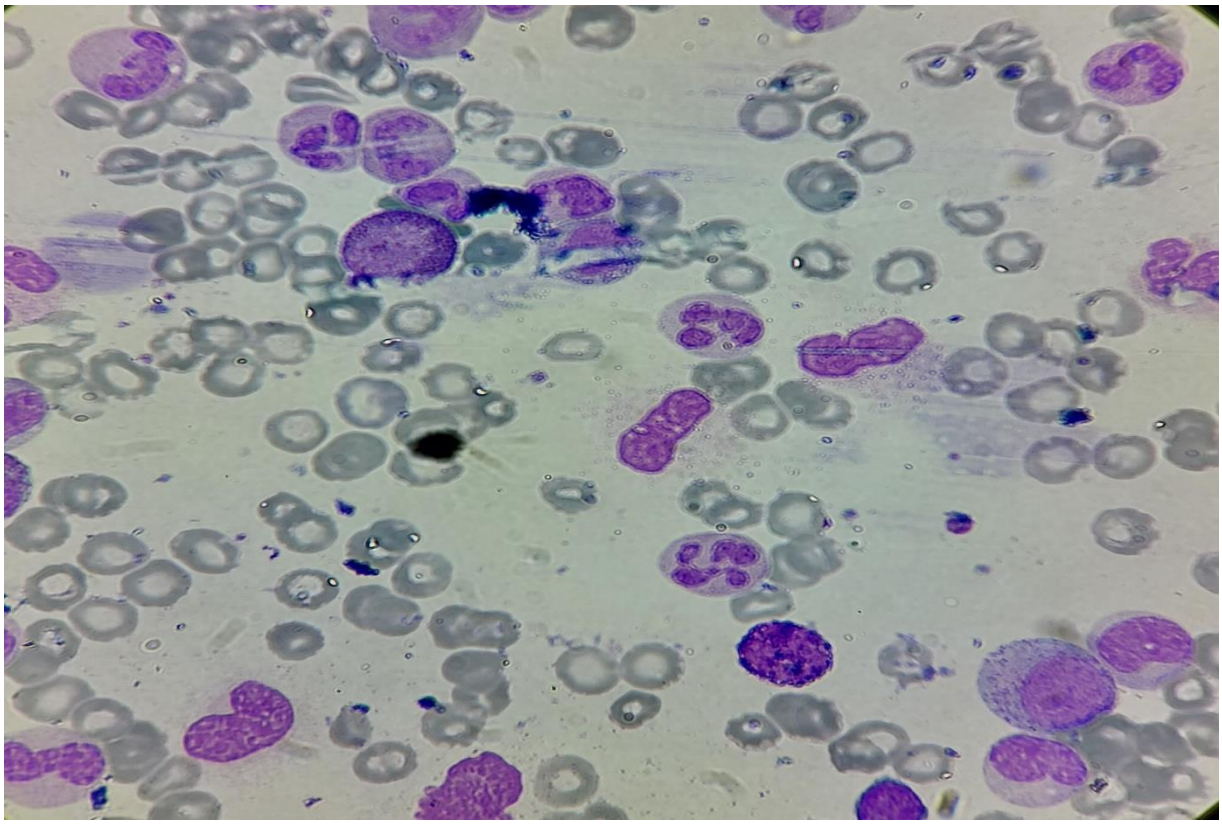


Figure 1,2 Photomicrograph of blood smear of patient showing marked leucocytosis with left shift of myeloid series and basophilia (Giemsa stain, x 400).

Discussion

Among all the childhood leukemias, chronic myeloid leukemia (CML), is a rare entity. Adult type of CML in children is even rarer and is characterized by Philadelphia chromosome [t (9:22)] positivity [4]. BCR-ABL fusion gene positive CML is better prognostic variant in paediatric age group and accounts for 3-5% of all childhood leukemias [5]. In recent years the incidence of leukemia in children and adolescents are on rise. The highest rise has been observed in the category of acute lymphoblastic leukemia. CML is very rare in this age group. CML in childhood presents as either of the two clinically distinct syndromes i.e. adult type CML (ACML) which is Philadelphia chromosome positive, and juvenile CML, also known as Juvenile Myelomonocytic Leukemia (JMML), which is Philadelphia chromosome – negative. Diagnosis of such

cases in chronic phase can be done by hematological investigations. CML occurring in children is called as adult form CML, which has the same clinical, morphologic and cytogenetic findings as adult type Ph positive CML. Clinical features of ACML are similar to that seen in CML occuring in adults. Hepatomegaly, splenomegaly and generalized lymphadenopathy, anemia, leukocytosis have been observed in all the patients (6). Three phases have been described for CML. Most patients are diagnosed in the first phase, called the chronic phase with median duration of 4-5 years. It can develop over time into the second-accelerated phase (6-8 months) and third- blast crisis phase (3-9 months). Accelerated and blast phase has worst prognosis(7). Allogenic bone marrow transplant is the most successful therapy if a suitable HLA identical donor is available for chronic phase CML . For patients without a suitable donor, control of the disease with chemotherapy (either hydroxyurea/busulphan or alpha interferon) is the best current alternative(5,8).

Conclusion

CML has a more aggressive clinical progression in paediatric age group. Treatment-free remission is often achieved in adults at present, but is not always applicable in pediatric CML. Early diagnosis and chemotherapy usually help in a better prognosis. Currently more researches are being conducted in this field.

Reference

1. Pushpam D, Bakhshi S. Pediatric chronic myeloid leukaemia: Is it really a different disease? Indian J Med Res. 2019;149: 600-9.

2. Hijiya N, Schultz KR, Metzler M, et al. Perspectives Pediatric chronic myeloid leukemia is a unique disease that requires a different approach. Blood. 2016;127:392-9.

3. Huho AN, Issaq N, Iacobas I, et al. A Rare Case of Pediatric Chronic Myelogenous Leukemia Presenting With Severe Thrombocytosis Without Leukocytosis. Pediatric and Developmental Pathology. 2018; 21:100-4.

4. Malhotra KP, Sharma CB, Jain J, Grover RK. Is b3a2 a better prognostic variant in childhood chronic myeloid leukemia? Journal of postgraduate medicine. 2010; 56 (3): 212-222.

5. Munoz A, Bureo E, Ortega JJ, Richard C, Olive T, et al. Treatment of ph-1 positive chronic myelogenous leukemia in children: Comparison between allogenic bone marrow transplantation and conventional chemotherapy. Haematologica 1988; 83: 981-84.

6. Mishra A, Tripathi K, Mohanty L and Pujari S. Philadelphia Chromosome positive chronic myeloid leukemia in a child: A case report. Indian Jr Hemat Blood Transfusion 2010; 26 (3): 109-110.

7. Rabinowitz I, Larson RS. Chronic myeloid leukemia. In: Greer JP, Foerster J, Lukens JN, Rodgers GM (edi). Wintrobes clinical hematology. Lippincott Williams and Williams. 11th edition. Volume 2. 2004: 2235-44.

8. Vardiman JW, Pierre R, Imbert M, et al. Juvenile myelomonocytic leukemia. In: Jaffe ES, Harris NL, Stein H, et al. Pathology and Genetics of Haematopoitic and Lymphoid Tissues. World Health Organization Classification of Tumors. Lyon, France: IARC Press; 2001:55-57.