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Case Report

Juvenile Chronic Myelomonocytic Leukemia - A Case Report

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ABSTRACT

Juvenile chronic myelomonocytic leukemia (JCMML) is a rare haemopoietic malignancy of early childhood. It represents upto 2% of haemopoietic malignancies in childhood. It has nevertheless formed the basis of fascinating biomedical research on oncogneic process in haemopoietic neoplasia over the past two decades along with insight into mutual relationship between inherited predisposition syndromes and myeloid leukemia. It is a stem cell disorder characterized by clonal hyperproliferation of monocytes and granulocytes without differentiation arrest. We present a case report of 6 years female child who presented with complaints of fever and pain in the abdomen

Key words: juvenile, myelomonocytic, myeloid leukemia.

INTRODUCTION

Juvenile chronic myelomonocytic leukemia (JCMML) is a rare haemopoietic malignancy of early childhood representing 2 to 3 % of all pediatric leukemia cases.[1] Myelodysplastic syndromes (MDS) in childhood are rare.[2] Their clinical and hematological features overlap with adult type MDS as well as myeloproliferative syndrome. Now WHO has clubbed all these entities into a category of myelodysplastic /myeloproliferative neoplasm.[3] It usually runs an aggressive clinical course with median duration of survival of children left untreated being less than 12 months from diagnosis.[4]

CASE REPORT

A six years old female child presented with fever of 2 months duration and pain in abdomen since eight days. Patient had no similar complaints in the past. Birth history was normal. Patient was fully immunized and had normal developmental milestones. On examination – General condition of the patient was stable with pulse 120/minutes. Respiration rate 26/minute, mild anemia. There was no sign of cyanosis, jaundice, petechia and skin rash. Patient had enlarged cervical, axillary and inguinal lymph nodes of size ranging between 1 to 3 cm. patient's height was 101cm, weight 13.5kg, head circumference 51cm, chest circumference 53cm. Spleen was palpable 3 cm, firm in consistency with well defined borders. Liver was palpable 5 cm below right costal

margin, firm in consistency. Rest of the systemic examination was unremarkable.

USG: abdomen showed hepatosplenomegaly.

CT scan: revealed bilateral axillary and mediastinal lymphadenopathy.

X-ray chest: no significant abnormality was seen.

Investigations

Hb-9.3%,TLC- 70800/cmm, neutrophils – 75%, lymphocytes- 3%, blastlike cells-6%, promyelocytes-6%, myelocytes-3%, metamyelocytes- 4%, monocytoid blasts and precursors-2%, monocytes-1%.

Peripheral blood smear: anisocytosis, moderate hypochromia, polychromatic RBCs with 10 nucleated RBCs per 100 WBCS. Leukocyets showed shift to left (Figure 3and 4)

Bone marrow: Myeloid hyperplasia with monocytoid precursors. Erythroid series showed normoblasts showing dysplastic changes and few megaloblasts. Few megakaryocytes showed hypolobulation. Haemolytic profile showed positive foetal fraction test. Sickling test was negative. (Figure 1 and 2)

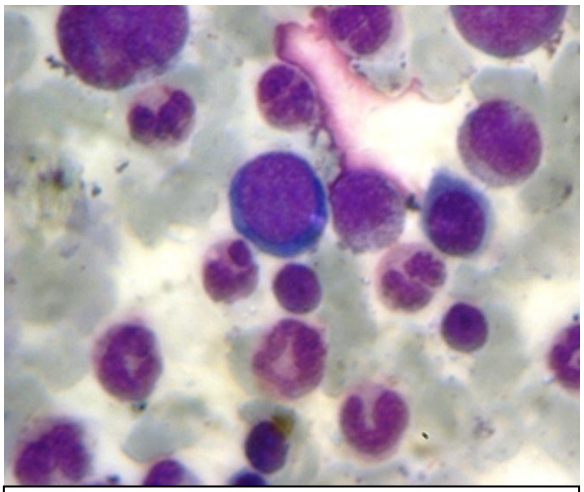

Fig1: Bone marrow showing myeloblast with precursor cells (40x,Leishman stain).

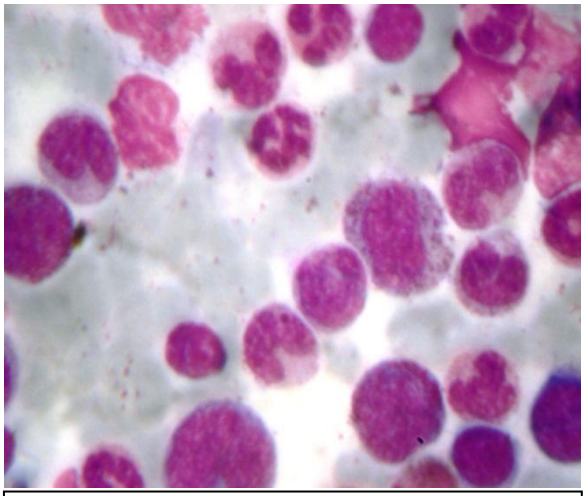

Fig2: Bone marrow showing myeloid and monocytic precursor cells(40x,Leishman stain).

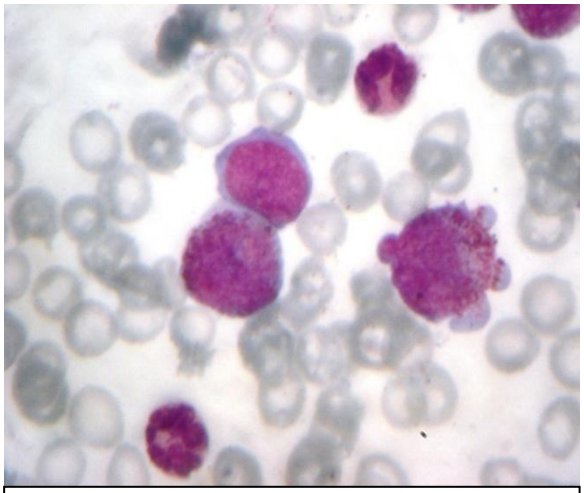

Fig3: PS showing myeloblast with precursor cells (40x,Leishman stain).

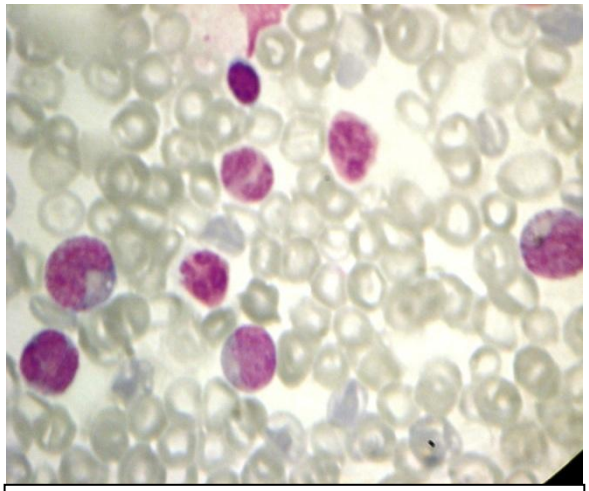

Fig4: PS showing monocyte with precursor cells (40x,Leishman stain).

Ancillary studies Hb F was 59.2%. by Bio-Rad HPLC method. LAP score was 7. Chromosomal analysis showed normal female karyotype 46xx. There was no numerical or sructural abnormality. Molecular cytogenetics (FISH) revealed no evidence of BCR/ABL, Ph chromosome, monosomy 7 or del (7q).

DISCUSSION

In 1976 the FAB co. op. group initially defined RABE and CMML as preleukemia. 6 years later FAB group added three more categories to this classification and adopted the preset term MDS.[5] WHO in their classification[3] of MDS define JCMML as myelodysplastic /myelo-proliferative disorder that encompasses the condition previously known as Juvenile myeloid leukemia and infantile monosomy 7 syndrome. JCMML is a chronic myeloproliferative disorder that typically affects young children. More than 95% cases are diagnosed before age 4 years. The chacteristic features include splenomegaly , leukocytosis with monocytosis more than 1000/cmm, thtrombocytopenia and anaemia, presence of immature myeloid precursors in peripheral blood. Bone marrow aspirate shows less than 20% blasts, high F heamoglobin level. No ph chromosome on cytogeneticc study and hypersensitivity to GMCSF. About 15% of cases are associated with Neurofibromatosis type I. many patients are asymptomatic with the diagnosis established on laboratory screenings.[6] The pathogenesis of JCMML arises from dysregulation of signal transduction through the RAS pathway. Infection remains usually the principle cause of death. It has bad prognosis late age of onset, decreased platelet count, high Hb F are bad prognostic signs. Bone marrow transplant (BMT) has been proposed as the treatment of choice for children with MDS However long term survival in patients of JCMML to receive allogenic BMT is only 30 percent which is not different than that in patients without BMT.[7] In the present case, clinical and laboratory findings (haemogram, PS and BM) were indicative of the diagnosis which was further substantiated by cytogenetic study for Ph chromosome and molecular cytogenetics (FISH). High HbF level (59.5%) was also in favour of the diagnosis.

CONCLUSION

Juvenile chronic myelomonocytic leukemia is rare childhood malignancy.Dignosis is on the basis of peripheral smear examination ,bone marrow examination and using ancillary studies like HbF,karyotyping,LAP score.Early diagnosis is important for treatment of these patient.

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