INTERNATIONAL JOURNAL OF MEDICAL AND APPLIED SCIENCES
E-ISSN:2320-3137

CASE REPORT

JUVENILE MYELOMONOCYTIC LEUKEMIA : A CASE REPORT

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Abstract :
Juvenile myelomonocytic leukemia (JMML) is a clonal hematopoietic disorder of childhood accounting for less than 2-3% of all leukemias in children. It was earlier considered to be a variant of chronic myeloid leukemia, but World Health Organisation (WHO) 2008 classification put it under the myelodysplastic/ myeloproliferative neoplasm. A five year old child was admitted under the provisional diagnosis of Protein Energy Malnutrition (PEM). He had a facial rash and presented with massive hepatosplenomegaly and cervical lymphadenopathy. On investigating, the child was found to have leucocytosis with precursor cells in peripheral blood; associated thrombocytopenia was also found. The bone marrow aspirate also showed increased number of precursors of granulocytes and monocytes. Fine Needle Aspiration Cytology (FNAC) from cervical lymph nodes and spleen showed infiltration by the leukemic cells. An increase in the fetal hemoglobin was detected.
Keywords: Fetal hemoglobin, Juvenile myelomonocytic leukemia, Myelodysplastic/myeloproliferative neoplasm.

INTRODUCTION

Juvenile Myelomonocytic Leukemia (JMML) , which is a clonal hematopoietic disorder characterized principally by the proliferation of granulocytic and monocytic series , presents commonly with features of respiratory involvement like pharyngo - tonsillitis or bronchitis. Fever, anaemia, hepatomegaly, marked splenomegaly, lymphadenopathy, an eczematous or maculopapular rash, xanthomas and a bleeding tendency are few other presenting features.[1,2,3]

Anaemia is present. Macrocytosis or microcytosis may be present depending on the associated genetic alterations. There is increase in the leucocyte count, together with neutrophilia and prominent monocytosis. Granulocyte precursors including blasts are often present. Some cases have eosinophilia or basophilia. Thrombocytopenia is usual and may be severe. The bone marrow is hypercellular with granulocytic hyperplasia and usually an increase in monocytes and their precursors, eosinophils or basophils. Megakaryocytes are often reduced in number. The blast percentage is often somewhat elevated. Trilineage dysplasia may be present.[1,2]

An altered Hb pattern is seen which is a readily accessible and reliable marker of JMML. At diagnosis, HbA_2 is reduced whereas HbF of more than 10% is found in two – third of the cases.[3,4]

JMML can progress to blast crisis, usually with French - American - British (FAB) M4 or M5 morphology, but more frequently succumb to disease due to tissue infiltration of myeloid cells.[4]

Case report : A 5 – year old boy presented with facial rash, massive hepatosplenomegaly, and cervical lymphadenopathy. His initial hematological investigations showed anaemia (Hb – 7.8gm%), leucocytosis (Total WBC count – 45,000/cumm) and severe thrombocytopenia (Platelet count - 15,000/cumm). The peripheral smear showed abnormal monocytic component (monoblasts and promonocytes accounting for 13%, monocyte count was 3%), and an increase of the granulocytic series. Consequently, a bone marrow aspiration was done and an increase of abnormal monocytic component (monoblasts and promonocytes accounting for 15%) was seen. FNAC from the cervical lymph nodes and spleen showed infiltration by leukemic cells, which was suggestive of the monocytic component. An initial screen by acid elution test showed an increase of the fetal hemoglobin which was later confirmed by hemoglobin electrophoresis. The child was then referred to a higher centre where cytogenetics was done , but no abnormal genetic alteration was detected.

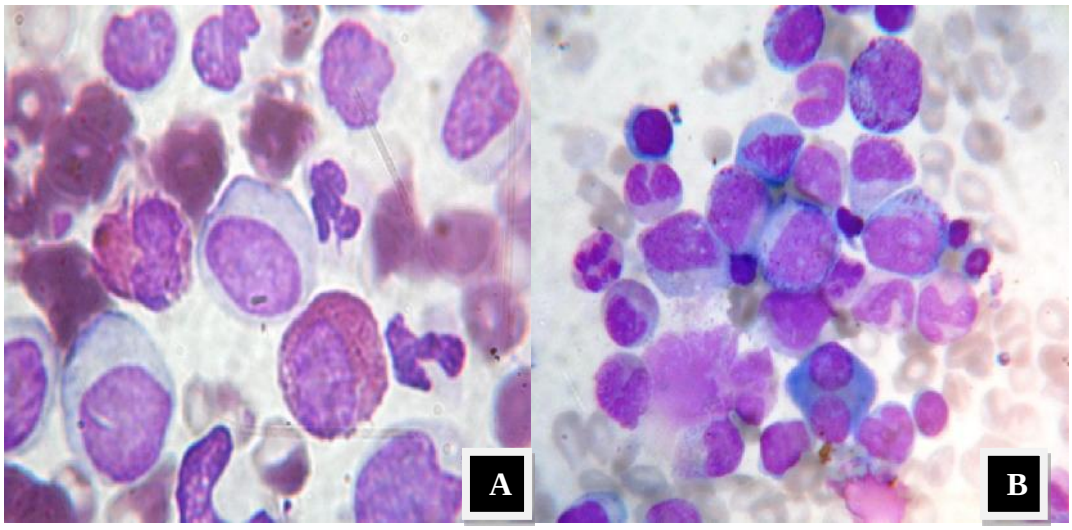
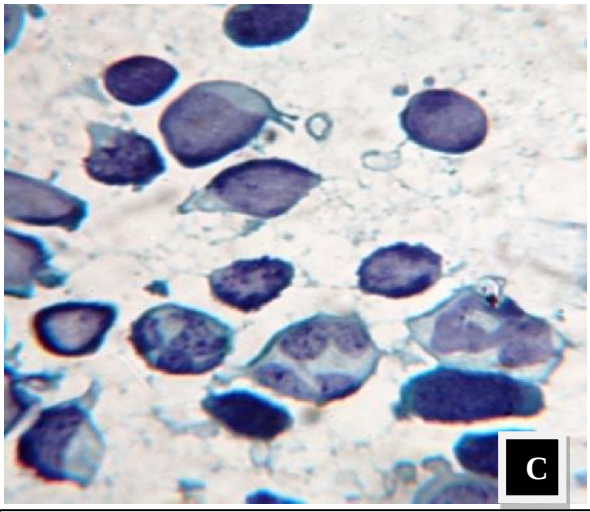


Fig: A – Peripheral smear showing increase of monocytic component,

B - Bone marrow aspirate showing the increase of granulocytic and monocytic component.



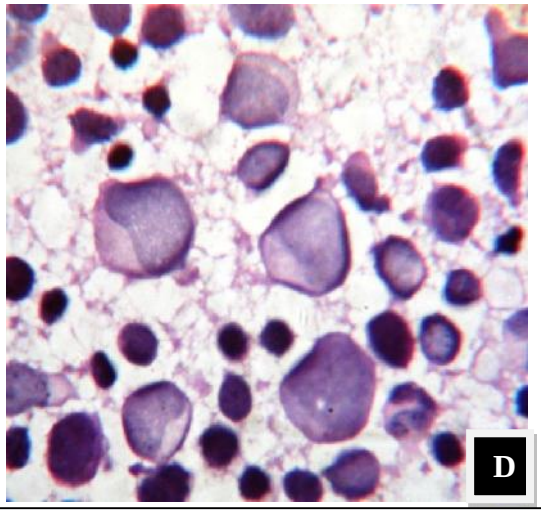


Fig: C – FNAC cervical lymph node showing infiltration by leukemic cells

Fig : D - FNAC from spleen showing infiltration by leukemic cells.

DISCUSSION

JMML, which was previously known as childhood chronic myeloid leukemia, has been put in the group of Myedysplastic / Myeloproliferative neoplasms by the 2008 WHO classification. The following criterion for diagnosis has been put forwarded by WHO [2]

(a) Monocyte count greater than 1×10^9/l
(b) Blasts plus promonocytes less than 20% in peripheral blood and bone marrow
(c) No Philadelphia chromosome or BCR - ABL1 fusion gene
(d) Two or more of the following

Haemoglobin F percentage increased for age

Immature granulocytes in the peripheral blood

White cell count greater than 10×10^9/l

Clonal chromosomal abnormality present

Myeloid progenitors hypersensitive to GM – CSF in vitro

The case under study had a monocytic count of 1350 / cumm with a high percentage of monocytic component (promonocytes and monoblasts) in both peripheral smear and bone marrow aspirate. Occasional dysplastic features like abnormal nuclear lobes, budding and abnormal mitosis was seen, justifying it to be a myelodysplastic / myeloproliferative neoplasm. Philadelphia chromosome, which was done at a higher centre, was not detected; HbF was increased, leucocytosis with immature granulocytes in peripheral smear was seen.

CONCLUSION

Any case with increased leucocyte count should be properly assessed with reference to the type of the precursor cells seen. In the present case, the monocytic component was overlooked at the initial count and a diagnosis of acute myeloid leukemia was thought of. FNAC proved to be an important tool in detecting the leukemic infiltration in the nodes and spleen.

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