

## Brief Reports

### Erythroleukemia: A Clinico-Hematological Review of Four Cases

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*Erythroleukemia is an uncommon disorder in children. Four cases of pediatric erythroleukemia, diagnosed over a period of nine years are presented. The patients presented with pallor, fever and hepatosplenomegaly of recent onset. Peripheral smear examination showed anemia, thrombocytopenia and circulating blasts. The bone marrow displayed erythroid hyperplasia with dysplasia and PAS positive erythroblasts. Myeloid blasts were myeloperoxidase positive and one case showed positivity for non specific esterase, indicating monocytoid differentiation, a poor prognostic feature. Prognosis was poor and follow up period was short.*

**Key words:** Erythroleukemia, Myeloid leukemia.

Erythroleukemia is a rare disorder characterized by uncontrolled proliferation of erythroblasts and myeloblasts comprising 2-7% of all acute myeloid leukemias. The French American British (FAB) group proposed the classification of these leukemias as AML M6(1). They are generally seen in old age(2). Very few cases of pediatric erythroleukemia have been reported in literature, comprising less than 1% of pediatric leukemias. A sixteen-year-old study at Memorial Sloan Kettering Cancer Center, New York reported only one case of pediatric erythroleukemia(3). Similarly, Day, *et al.*(4) published a report of two cases of erythroleukemia in infancy.

We present four cases of erythroleukemia in the pediatric age group diagnosed at our unit over a period of nine years.

#### Subjects and Methods

Hemogram, bone marrow findings and

relevant clinical features of 297 patients of acute myeloid leukemia, seen in the department of Hematology at the All India Institute of Medical Sciences, New Delhi between January 1990 to December 1998 were reviewed. Twenty-one cases of erythroleukaemia were diagnosed. Of these, four were children and 17 adults. The initial work-up included a history elicitation, examination and necessary biochemical investigations. Complete hemogram, peripheral smear, bone marrow examination and cytochemistry including Sudan Black, non-specific esterase, acid phosphatase and periodic acid Schiff were performed in all cases. The FAB criteria of presence of >50% erythroid precursors and >30% non-erythroid blasts in the bone marrow, was followed for diagnosis of acute erythroleukemia [AML (M6)](1).

#### Results

The comparative evaluation of the

presenting clinical features in children and adults is presented in *Table I*. The patient's age ranged from 8 years to 75 years with a mean of 35.4 years. Of these, four patients were in the pediatric age group. The male female ratio was 3:1 and 1:1 in the pediatric and adult population respectively.

The four pediatric patients are described below:

*Case I:* A ten-year-old female presented with fever and pallor for six months with sternal tenderness, cervical lymphadenopathy and congestive heart failure. On examination, there was marked pallor (Hb 2.3 g/dL) with thrombocytopenia. Blasts were present in circulation.

*Case II:* An eleven-year-old male presented with high-grade fever and epistaxis for 15 days. On examination he had pallor, sternal tenderness and mild hepatomegaly. Lymph nodes were not enlarged. Laboratory investigations showed pancytopenia. No blasts or nucleated red cells were present in circulation.

*Case III:* A twelve-year-old male presented with low grade fever and progressive pallor for five months. On examination, he

showed mild gum hypertrophy, generalized lymphadenopathy and hepatosplenomegaly. Laboratory investigations showed anemia (Hemoglobin 5.6 g/dL), thrombocytopenia and a raised total leucocyte count with circulating blasts and nucleated red cells.

*Case IV:* An eight-year-old male presented with progressive pallor for 11 months. There was no organomegaly or lymphadenopathy. The peripheral smear showed pancytopenia and nucleated red cells. No blasts were seen in circulation.

Bone marrow aspirates were cellular in all cases. They displayed a prominent erythroid hyperplasia, erythroid precursors forming more than 50% of the cell population, with features of dyserythropoiesis like megaloblastoid erythroblasts, multinuclearity and nuclear budding and bridging. The mean myeloblast population was 27.6% in the pediatric patient group. Auer rods were found in one case (Case I).

Special stains were performed in all the cases namely myeloperoxidase/Sudan Black, non specific esterase and periodic acid Schiff. Three of the four cases were positive for Myeloperoxidase /Sudan Black. In one case

**TABLE I—Comparison of Presenting Clinical Features in Pediatric and Adult Patients of AML -M6.**

Characteristics	Age <12 years	Age >12 years
Number	4	17
Sex: Male	3	11
Female	1	6
Mean duration of presenting symptoms	5.75 months	5.69 months
Hepatomegaly	2 (50%)	11 (64.7%)
Splenomegaly	1 (25%)	7 (41.2%)
Jaundice	None	one
Bleeding manifestations	1(25%)	3 (17.6%)
Generalised lymphadenopathy	1(25%)	None

### Key Messages

- Pediatric erythroleukemia is an acute onset leukemia.
- Clinical profile and hematological features are similar to adults.

the blasts were also positive for non-specific esterase, indicating myelo monocytic differentiation, a poor prognostic feature. Erythroblasts showed positivity for periodic acid Schiff in two cases only. Genetic analysis for chromosomal abnormalities could not be performed.

#### Discussion

Erythroleukaemia is a rare disorder comprising of 2-7% of cases of acute myeloid leukemia. In our study, 21(7.1%) out of 297 cases of AML were diagnosed to have erythroleukemia over a period of nine years. Pediatric erythroleukemia is very rare. Only small number of cases of erythroleukemia have been diagnosed world wide in the pediatric population, most of which are case reports(3-5). Literature regarding the clinico-hematological profile in children is not greatly available, especially in the Indian population.

In our study, most pediatric patients presented with pallor, fever and hepatosplenomegaly of recent onset. Severe anemia was noted associated with thrombocytopenia in the peripheral smear with circulating blasts and nucleated red blood cells. The bone marrow was cellular with erythroid hyperplasia. Erythroid precursors showed megaloblastosis, dyserythropoiesis and bi and tri nucleate precursors. Myeloid blasts were Sudan Black/ Myeloperoxidase and PAS positive. One case showed Auer rods.

One out of the 4 (25%) cases showed the presence of non-specific esterase positivity in the blasts, thus indicating monoblastic

differentiation. This is a possible indicator of adverse prognosis. Secondary AML or AML arising on a prior background of MDS was not seen in any of our patients. Other indicators of poor prognosis include presence of splenomegaly at the time of diagnosis, monoblastic differentiation of myeloid blasts and the presence of Auer rods in the myeloblasts.

These patients were induced with a classical regime of Injection cytosar 100 mg/m<sup>2</sup>/day 1-7 IV Injection daunorubicin 45mg/m<sup>2</sup>/day on day 1-3 I.V. and oral prednisolone. Three or more cycles of induction were required. This was followed by maintenance on high dose cytosar 3g/m<sup>2</sup> BD on alternate days for three such doses *i.e.*, on day 1, 2 and 5. Response to treatment was poor in most cases. The mean follow-up period was only 3 months with most patients refusing therapy after prognosis was explained. Only one patient achieved remission after two cycles of induction therapy after which he was lost to follow-up.

It is thus concluded that pediatric patients of AML (M6) present with a short duration of illness. Most common presenting features are pallor and fever. Examination findings and hematological findings on peripheral smear and bone marrow are similar to classic cases of AML M6 in adults. Prognostication could not be done in this study due to small number of patients and short duration of follow up.

*Contributors:* RS designed the study; AKS collected the data; VVB collected data, performed statistical

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analysis and will act as guarantor of the paper.

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## Rotavirus Infection in Children with Acute Diarrhea as Detected by Latex Agglutination, ELISA and Polyacrylamide Gel Electrophoresis

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*We examined prospectively, stool specimens from 135 children, 0 to 3 years old, referred for fever, abdominal pain, vomiting and/or acute diarrhea. Rotavirus antigens were detected from fecal samples by latex agglutination (LA), ELISA and polyacrylamide gel electrophoresis (PAGE). Rotavirus antigen positivity by Latex, ELISA and PAGE were 15, 55%, 12.59% and 11.85%, respectively. With PAGE test as reference, the sensitivity and specificity of LA and ELISA tests was 93.75%, 94.96% and 100%, 99.16%, respectively, The positivity ratio between 13-24 months group was meaningful with all tests ( $P = 0.042$  for LA;  $P = 0.05$  for ELISA;  $P = 0.031$  for PAGE). ELISA and LA use found to be as sensitive and specific as PAGE in the diagnosis of rotavirus diarrhea.*

**Key words:** Diarrhea, Gastroenteritis, Rotavirus.

Gastroenteritis is a leading disease causing death among infants in developing countries(1). In our country, the incidence in rural area is reported to be 28% and increases