

## Severe Pallor In Neonate –Case Series Of Congenital Leukemia

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**Abstract:** 3 neonates presented with severe pallor in early gestational period. Complete blood count with peripheral smear examination showed abnormal cells suggestive of Congenital Leukemia which was a Rare diagnosis. Bone Marrow Examination confirmed the Diagnosis. All patients needed blood transfusion. However, Immunohistochemistry and Cytogenetic study could not be done due to paucity of investigation in our institute and patients were referred to higher centre.

**Key Words:** Neonates, Pallor, Leukemia.

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### I. Introduction:

Congenital leukemia is luckily a very rare hematologic malignancy that originate in utero and usually get diagnosed from birth to 6 weeks of life. The annual incidence estimates to about 1-5 per million live births. Although the etiology is unknown, the presence of leukemia at birth suggests genetic abnormalities and possibly intrauterine exposures to drugs or other toxins as contributing factors.<sup>1,2</sup>

The criteria for diagnosis of congenital leukemia are:

- a) Disease presentation at or shortly after birth (within 30 days of birth).
- b) Proliferation of immature/ precursor white cell population.
- c) Infiltration of cells into extra-haemopoietic tissues
- d) Absence of Congenital leukemia simulators.<sup>3,4</sup>

Congenital leukemia simulators include TORCH infection, Congenital HIV Infection, Hemolytic disease of the newborn (ABO or Rh incompatibility), hereditary spherocytosis, twin-twin transfusion, other neoplastic infiltrates (metastatic neuroblastoma, rhabdomyosarcoma, Langerhans cell histiocytosis). Neonatal or congenital leukemias are generally myeloid in origin unlike childhood leukemia which are lymphoid in origin. Neonatal leukemia is more likely to present with poor prognostic factors. Neonatal ALL has a disease-free survival rate of ~10% compared to >70% in older children<sup>5</sup>. Neonatal leukemia is the leading cause of death in neonate due to neoplastic disease. There is high index of suspicion in cases of Down's syndrome and hydrops fetalis. Essential karyotyping in such individuals antenatally by collecting fetal cells through amniocentesis or cordocentesis should be done for cytogenetic study and demonstration of blast forms.<sup>6,7</sup>

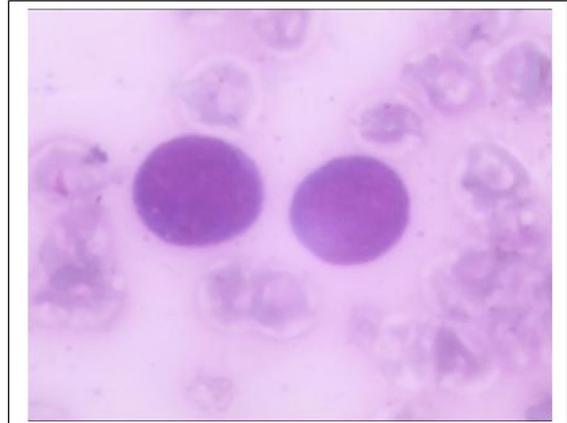
### II. Case Series:

#### Case 1:

The girl baby (27 day old) born out of of an uneventful pregnancy, G<sub>1</sub>P<sub>0</sub> with no history of consanguinity, normal vaginal delivery, birth weight 2.9kg. Baby presented with refusal to suck and gradual swelling of abdomen for last 15 days, fever for last 10 days. On clinical examination, vitals were stable with no features of shock. Baby had severe pallor with multiple purpuric spots and red nodules (Leukemia cutis) over skin. (Table/Fig 1.) Spleen was palpable 5cm below costal margin. CBC showed marked leukocytosis over 1 lakh (110000/cmm), low platelet count (35,000/cmm) and reduced haemoglobin (6 gm%) and atypical cells. LFT showed conjugated hyper bilirubinemia (total serum bilirubin-13.12mg/dl, conjugated 8.5 mg/dl). Torch screen-negative. Usg whole abdomen- splenomegaly (9.2cm) with mild ascites. Bone marrow biopsy showed- immature myeloid cells (meta, myelo promyelocytes) 48% and Blast cells-30%. (Table/Fig 2)



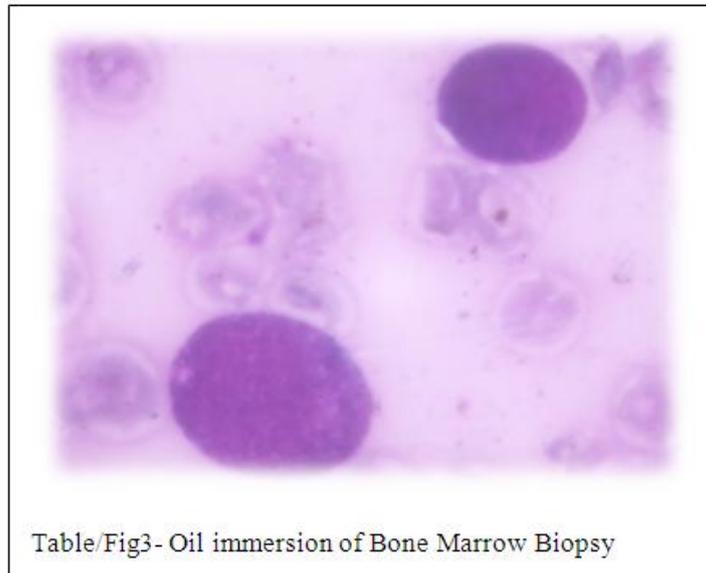
Table/Fig 1-Leukemia Cutis



Table/Fig-2-Bone Marrow Biopsy showing Blast Cells.

**Case 2:**

36 wks,preterm,Small for gestational age baby,Born out of non-consanguinous marriage,delivered by Caesarian Section due to Premature rupture of membrane,birth weight 1.9kg, presented with lethargy and paleness since birth.On physical examination at day 7 of life,skin had multiple purpuric spots.Vitals were stable.Baby had occasional episodes of hypoglycemia with liver palpable 4cm below costal margin. CBC showed decreased leukocyte count(2000/cmm), low platelet count(10,000/cmm) and reduced hemoglobin (6 gm%) and atypical mononuclear cells -26%.Band cells-4%.LFT showed conjugated hyper bilirubinemia( total serum bilirubin-10.23mg/dl, conjugated 7.5 mg/dl).Torch screen- negative.USG whole abdomen- significant ascites with hepatomegaly.Urine for AFB detection RNA PCR-Negative.Bone marrow biopsy showed - Blast cells in increased number constituting about 25% of the marrow nucleated cell population. These blast cells are largely pleomorphic having dispersed chromatin, mainly of myeloid origin, suggestive of acute leukemia.(Table/Fig3)



Table/Fig3- Oil immersion of Bone Marrow Biopsy

**Case 3-**

Term,Appropriate for gestational age boy baby,born from an uncomplicated normal vaginal delivery presented on day 27 of life with multiple echymosis(Table/fig 4) and red nodules (Table/fig 5)all over the body.Patient was apparently well 7 days back and gave no history of fever.On physical examination patient had severe pallor.Head to foot examination revealed mongoloid slant of eyes,sandle foot gap and simian crease.Vitals were stable and patient was feeding well.Liver was palpable 5 cm below costal margin and spleen was palpable 4cm below costal margin.CBC showed marked leukocytosis(64000/cumm) ,low platelet count(45,000) and reduced haemoglobin(5gm%).Peripheral smear examination showed marked immature myeloid cells and myeloblasts (74%).Rbc was normocytic,normochromic.Bone marrow aspiration from tibial

tuberosity showed hypercellular marrow with myeloid maturation shift to the left with increased myeloblast constituting about 30% of the marrow non erythropoid cell population. Megakaryocytes and erythropoiesis was markedly depressed. LFT showed conjugated hyper bilirubinemia .TORCH screen and blood culture was negative. Karyotyping showed triple trisomy 21.

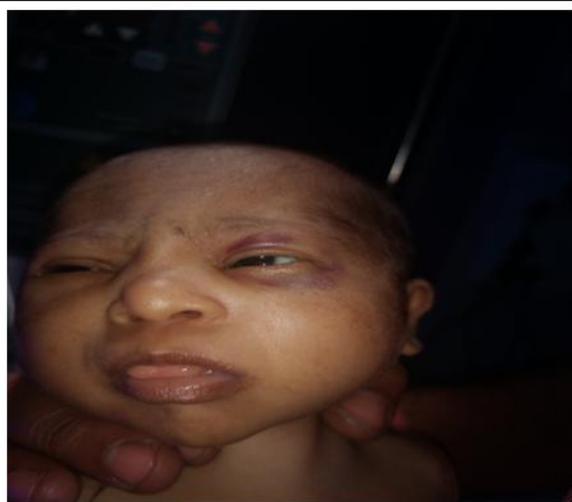


Fig 4-Ecchymosis over left eyelid.



Table/Fig 5-Red nodules on back

### III. Discussion:

All the three cases were initially thought to be late onset sepsis and were managed with IV fluids and antibiotics. Blood transfusion was given. TORCH Screening and blood culture was sent for all three patients which came to be negative. There was no improvement in blood picture after repeated transfusion. The counts showed pancytopenia in one patient with Leucocytosis in the other two. Peripheral Blood Examination showed Abnormal Cells in all three patients and Bone Marrow examination confirmed the diagnosis. Patients were referred to higher centre for further management.

### IV. Conclusion:

Although congenital Leukemia is a rare diagnosis, it should be considered as a possibility in all newborns who do not respond to conservative management. This will help in early diagnosis, prompt management and less mortality.

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