International Journal of Advanced Research and Review

www.ijarr.in

# Congenital Acute Leukaemia - A Case Report

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## ABSTRACT

Acute leukaemia is rare in neonates. Only a few cases have been reported so far. Congenital leukaemia occurs at a rate of 1 per 5 million births. The majority are Non-Lymphocytic type (80%), while acute lymphoblastic leukemia (ALL) comprises only < 20%. Familial neonatal leukemia is extremely rare and no child born to a mother with leukemia has been found to have the disease during the neonatal period. We are reporting here a rare case of lymphomonocytoid type of congenital leukaemia in a newborn.

**Key Words-:** Congenital Leukaemia, Newborn Infant

## INTRODUCTION

Acute leukaemia is rare in infants. Only a few cases have been reported so far. Congenital leukaemia occurs at a rate of 1 per 5 million births. It is characterised by non specific symptoms requiring a high index of suspicion for further investigations and diagnosis. Congenital and neonatal leukemia occur rarely, yet carry high mortality rates and pose special problems for the perinatologist and hematologist. Althrough 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.

## CASE REPORT

We are reporting here a rare case of lymphomonocytoid type of congenital leukaemia in a newborn. Baby was full term with no antenatal history of maternal fever or rash, delivered by normal vaginal delivery with birth weigth 2.8kg was admitted in NICU of Umaid Hospital, Jodhpur. Baby was having purpuric spots over face and multiple petechiae over chest, abdomen and back. Liver was 2cm below costal margin and Spleen was palpable 3 cm below costal margin. On 2$^{nd}$ day of life baby developed bleeding per rectum. There was no history of maternal exposure to radiation or drugs except IFA tab. Maternal age was 30 years at the time of conception.

On examination baby was sick, pale with purpuric spots over face at dorsum of nose and at below right eye. Petechial spots were present over chest, abdomen and back. There was no evidence of microcephaly, cataract, facial dysmorphism, lymphadenopathy. Liver was palpable 2 cm below costal margin and spleen was present 3 cm below costal margin. External genitalia were normal. Auscultation over chest was normal. Baby had normal pulses and normal capillary filling time. Heart sounds were normal. There was no focal neurological deficit.

Investigations revealed Hb-17.6gm%, TLC-340000/cumm with Polymorphs-06%, Lymphocytes-23%, Monocytes-02%, Eosinophis and Basophils-0% and Lymphomonocytoid cells forms 69%. Peripheral blood smear showed normocytic normochromic RBC, moderate polychromasia, Nucleated RBC 11/100 WBC (fig.1). There was marked leucocytosis, many blast cells were having lymphomonocytoid configuration. Many smudge cells were seen. Lymphomonocytoid cells formed 69% of total cell population. Platelets showed marked thrombocytopenia with no hemoparasites were seen. Peripheral Blood Smear was suggestive of congenital acute leukaemia. Urinanalysis and Chest radiograph was normal. Ultrasound scan of abdomen showed hepatosplenomegaly. Blood urea and Serum creatinine was in normal limits. Bone marrow examination revealed same finding as that of peripheral blood smear which was suggestive of congenital acute leukaemia

Baby was admitted to NICU of Umaid Hospital just after birth and started intravenous fluids and antibiotics. Baby had bleeding per rectum on day 2 & 3$^{rd}$ day of life, for that appropriate treatment was given. Baby deteriorated for first 7 days than started improving and discharged against medical advice on 15$^{th}$ day of life.

## DISCUSSION

Congenital leukemia is a very rare disorder[1]. Only 200 reports of congenital leukemia are published in literature[2]. The majority are Non-Lymphocytic type (80%), while acute lymphoblastic leukemia (ALL) comprises only < 20%. Familial neonatal leukemia is extremely rare and no child born to a mother with leukemia has been found to have the disease during the neonatal period[3]. Congenital leukemia is occasionally associated with number of congenital anomalies and with chromosomal disorders such as Down's syndrome, Edward's and Patau's syndrome and a number of nonspecific chromosomal abnormalities which was not seen with our case. Subtle cytogenetic abnormalities may occur more commonly in the affected infants and their parents, when studied with newer cytogenetic techniques[4].

The clinical signs of leukemia may be evident at birth with hepatosplenomegaly, petechiae and ecchymosis. Leukemic cell infiltration into the skin (leukemia cutis) is commonly found[3]. In infants in whom the disease develops within the first month (not at birth), the symptoms are ill defined with low-grade fever, diarrhea, hepatomegaly and failure to gain weight. Leukemia cutis is less common[3]. Clinically, it is important to differentiate congenital leukemia from other leukoerythroblastic conditions, which are seen in response to bacterial infection, hypoxemia and severe hemolysis in the neonate[5]. Other differential diagnosis includes congenital syphilis, intrauterine viral disease, neuroblastoma and the transient myeloproliferation syndrome associated with Down's syndrome [6,7,8]. In our case patient has improved with only symptomatic treatment and diagnosis was confirmed by PBF and Bone marrow findings.

Cellular morphology, immunophenotype and chromosomal studies differentiate acute lymphoblastic from acute non lymphoblastic leukemia found in newborns[4]. FAB classification based on cell morphology reveals that the most common subtype in infantile and neonatal acute nonlymphocytic leukemia is the monocytic variety[9,10]. The most common locus involved in translocation in infantile acute lymphoblastic leukemia is at 11q 23 this is involved in at least

50% of infant leukemias and in many neonatal cases[11]. We are unable to do all above investigations as these are very costly and patient's parents can afford them. They are also not available at our centre.

The course of congenital leukemia is one of rapid deterioration and death from hemorrhage and infection. Specifically, it is a more aggressive disease with increased incidence of leukocytosis, massive hepatosplenomegaly, CNS involvement, thrombocytopenia, hypo-gammaglobenimia, disseminated intravascular coagulopathy (DIC) and less frequent remission induction by 14 days[12]. The current improved success of remission induction with treatment of acute non-lymphocytic leukemia in infants younger than 1 year is similar to that in older children using combination chemotherapy. However, the experience with newborns is limited, but between 1984 to 1989, 5 of 12 newborns with acute non-lymphocytic leukemia sustained complete remission with chemotherapy, and all were in the myelomonocytic or monocytic category [9,11]. However, in acute lymphoblastic leukemia, the treatment outcome is significantly poorer in infants younger than 1 year at diagnosis (23% disease-free survival compared with 70% for older children) and may be even lower in newborns. Only 10 to 20% survival for infants younger than 6 months of age at diagnosis has been reported as compared to 40% for those, older than 6 months[12,13]. Fewer than 15% of newborns with acute lymphoblastic leukemia have remission lasting more than few months. Success of Remission induction in Congenital AML is almost similar to that in older children using combination chemotherapy.

Indications for Post mortem diagnosis of acute congenital leukaemia were overt major and multiple minor congenital anomalies with facial dysmorphism not indicative of commonly seen genetic disorders.Suspected Inborn errors of metabolism.Suspected hematological / oncological disordersDiagnosis not confirmed during the life of the child[14] .

Investigations which can be done if death is inevitable & diagnosis not confirmed during the life of the child are Chromosomal Karyotyping, whole blood for DNA analysis, Urine for Microscopy, Biochemistry and metabolic screening, Bone marrow study (aspiration & biopsy) and Biopsies of Liver, Lung, Skin, Kidney[15].

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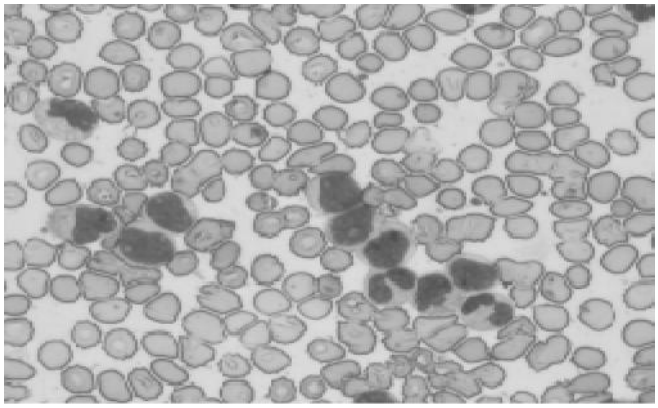

Fig 1: peripheral smear showing blasts and neutrophils