CASE REPORT

Congenital Leukemia

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Abstract

Congenital leukemia is a rare disease that can manifest soon after birth. Cutaneous involvement consists of red, brown or purple papules or nodules and purpura. We present a case of congenital myelomonblastic leukemia in a seven week old infant who had petechiae and subcutaneous nodules. Diagnosis was established by the presence of leukemic cells in bone marrow and involved skin along with cytochemical characterization of these cells.

Key Words

Congenital Leukemia, Congenital Myelomonoblastic Leukemia, Leukemia Cutis, Chloromas

Introduction

Congenital leukemia is defined as leukemia that develops in utero (1). Because of the doubling time of leukemic cells, the disease becomes clinically evident after birth or shortly thereafter (2). It is a very rare disease and is seen one per five million births (3). A large proportion of congenital leukemias are of myeloid lineage in contrast to paediatric leukemias in general, which are primarily lymphoid. Congenital leukemia is characterised by non specific symptoms which require a high index of suspicion for further investigations and diagnosis. We report a case of congenital myelomonobalstic leukemia with cutaneous involvement.

Case Report

A seven weeks old boy presented to paediatrics outdoor clinic with complaints of high grade fever and cough for five days along with irritability and excessive crying. The antenatal and early postnatal history was uneventful. The delivery was conducted by a competent gynaecologist in a government hospital. The child had normal APGAR score and birth weight of 2.75 kg. The immunization was complete till date. Shortly after admission the child developed pneumonia and respiratory symptoms. On examination, there were multiple well defined round to oval, firm, mobile, nontender, warm and erythematous nodules over the scalp and forehead. The overlying skin was smooth and shiny. There were ecchymotic spots over the whole body. The child was admitted for investigations. The haematological investigations revealed 5 gms/dl haemoglobin, 46,000/mm3 total leukocyte count and 40,000/mm3 platelet count. The peripheral blood smear showed normocytic normochromic blood picture with markedly reduced platelets and many blasts (*Fig.1*). The bone marrow aspiration showed increased M: E ratio with diffuse sheets of blasts. The blasts were large in size with cytoplasmic budding, nuclear infoldings, fine nuclear chromatin and 2-3 indistinct nucleoli (*Fig.2*). The cytochemistry was done and it turned out to be positive for Myeloperoxidase (MPO) (*Fig.3*), Chloracetate esterase (CAE) and Nonspecific Esterse (NSE) (*Fig.4*) and negative for Periodic Acid Schiff (PAS). So the case was classified as AML-M4 i.e. Myelomonoblastic leukemia (FAB). The chest x-ray of the patient revealed bilateral lung infiltrates. FNAC & histopathology of the skin lesions showed chloromas. Patient was referred to PGIMER Chandigarh and was under treatment for one year and responded to treatment.

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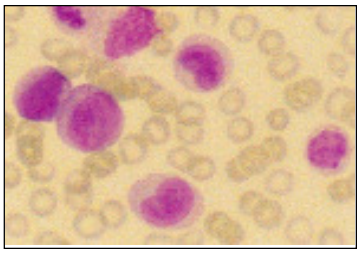

Fig.1 Peripheral Blood Film Showing Blasts (AML-M4)

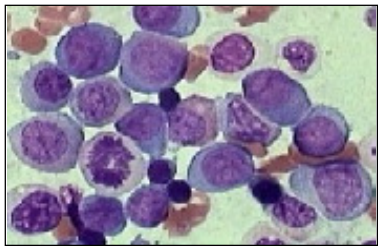

Fig. 2 Bone Marrow Showing Blasts (AML-M4)

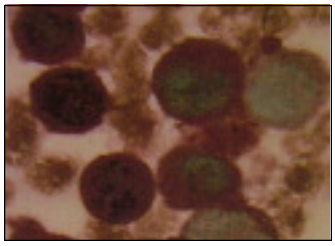

Fig. 3 Blasts Showing Positive Reaction to Non Specific Esterase (NSE) Stain (x1000)

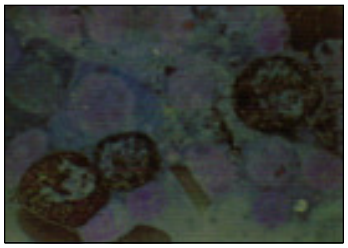

Fig. 4 Some of the Blasts Showing PositiveReaction to Myeloperoxidase (MPO) stain (x1000)

Discussion

Congenital leukemia is a rare disorder that usually presents with extramedullary infiltrates and myeloid phenotype in 80 % of cases (4). Leukemia cutis occurs in 25% -30% of infants with congenital leukemia and is more frequently associated with acute myeloid leukemia as compared to acute lymphoblastic leukemia (5). These patients usually present with petechiae, purpura, hepatosplenomegaly, lethargy, fever and pallor. Many infants die of respiratory distress secondary to pulmonary leukostasis and bronchopneumonia (1). One of the important physical manifestations of congenital leukemia is chloromas, which manifest as nodular infiltration of the skin. Chloromas are solid collections of myeloblasts. They are most common in the skin where these are termed as leukemia cutis (6). They are most commonly seen in M-4 and M-5 type of AML (7). Cutaneous involvement is usually in the form of red, brown or purple papules and nodules (blue berry -muffin lesions) (8,9). The diagnosis is established by presence of leukemic cells in biopsy specimens of the involved skin.

Association of chromosomal anomalies like Down's syndrome with congenital leukemia has been well documented. It is important to differentiate congenital leukemia from other leukoerythroblastic conditions which are seen in response to bacterial infections, hypoxemia and severe haemolysis in the neonates. Other differential diagnoses are congenital syphilis, intrauterine viral disease, neuroblastoma and transient myeloproliferative syndrome associated with Down's syndrome. Survival time in congenital leukemia is brief (1). Therefore, it is essential to differentiate congenital leukemia from myeloproliferative disorders associated with Down's syndrome which show a characteristic, complete clinical and haematological recovery within weeks or months of diagnosis without anti leukemic treatment.

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