CONGENITAL LEUKEMIA CUTIS

H J Shroff, H M Siddiqui, D A Parikh, R G Torsekar, H N Nandwani and U L Wagholikar

A case of congenital leukemia cutis in a female child starting at birth and manifesting as erythematous papules and nodules all over the body, along with hepato-splenomegaly and terminating fatally is reported. This is an extremely rare disease.

Key words : Congenital leukemia cutis, Acute monomyeloid leukemia (M4).

Cutaneous manifestations during the course of the disease have been reported in several types of leukemia. Congenital leukemia cutis (CLC), seen in the neonatal period, is a rare form of presentation. Skin manifestation is the first sign of the disease in a majority of the cases. Nearly, 150 cases of CLC have been reported in the world literature.[1] A case of CLC in a neonate, believed to be the first case of CLC is being reported from India.

Case Report

A 24-day-old Muslim female child, born to non-consanguinous parents as the first full-term normal delivery with 2.5 kg birth-weight, and having a generalised erythematous skin rash, was admitted in the hospital in May, 1983. On the first day after birth, the mother noticed a few small erythematous papules around the nose of the child and thought it to be insignificant. Subsequently, fresh lesions appeared which gradually spread all over the body including the scalp, palms and soles. The mother had no previous history of abortion, still-birth, premature delivery or consumption of any drug during this pregnancy. There was no history of any hematological disorder in the family. Father had suffered from syphilis in the past for which he was adequately treated. The child had moderate pallor but no icterus or petechial haemorrhages. Liver and spleen were enlarged by 3 cm and 2 cm respectively below the costal margins and were smooth, firm and non-tender. There was no lymphadenopathy. The cutaneous lesions consisted of multiple, discrete, bilaterally symmetrical, erythematous papulo-nodular eruptions present on the face, neck, trunk and extremities. Congenital syphilis, systemic mastocytosis, histiocytosis-X and erythroblastosis foetalis were considered in the differential diagnosis.

Laboratory investigations at the time of admission revealed haemoglobin, 8 gm/dl; leucocyte count, 2,50,000/cmm; platelet count, 26,000/cmm; ESR, 40 mm; peripheral blood smear showed myeloblasts 81%; promyelocytes, 8%; myelocytes, 3%; band forms, 2% and lymphocytes, 6%. There were no neutrophils,

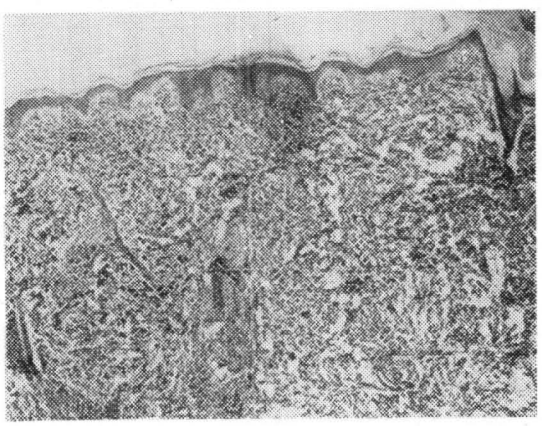

Fig. 1. A dense, diffuse infiltrate of leukemic cells involving the entire dermis and extending upto the sub-cutis. (Haematoxylin and Eosin x 250).

From the Departments of Dermatology & STD and Pathology, Grant Medical College & JJ Group of Hospitals, Bombay-400 008, India.

Address correspondence to : Dr. H.J. Shroff, Aboobakar Building, Near Regal Cinema, Colaba Causeway, Bombay-400 039, India.

eosinophils or basophils. A number of myeloblasts showed monomyeloid features; Auer rods were seen in some. A smear of the bone marrow revealed marked hypercellularity owing to a heavy infiltrate of myeloblasts and other immature myeloid cells. The normal marrow elements were severely depleted. The immature myeloid cells in the blood and bone marrow showed good positivity with myeloperoxidase and Sudan black staining procedures. The blast cells, however, did not stain with the PAS technique. Roentgenograms of the chest and the skull were normal. Histopathologic examination of an erythematous skin nodule on H & E staining showed thinning of the epidermis and a dense infiltrate of myeloblasts and other immature leukemic cells in the dermis extending from the epidermis to the sub-cutis. The adnexa were completely enveloped by the leukemic cells (Figs. 1 and 2). The above

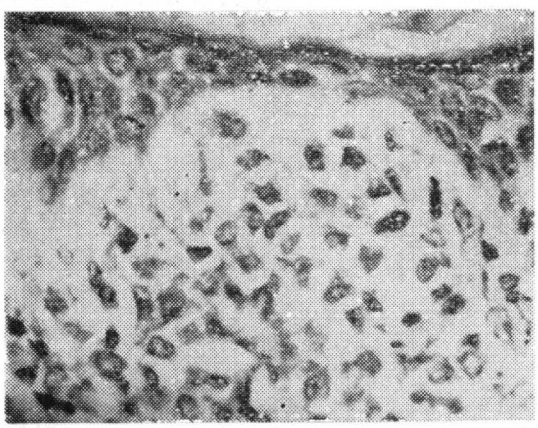

Fig. 2. Atrophic epidermis with an infiltrate of leukemic myeloblasts in the uper dermis. The tumor cells have large round or indented, slightly pleomorphic and hyper-chromatic nuclei, with one or more nucleoli. (Haematoxylin and Eosin x 1000).

findings confirmed the diagnosis of congenital leukemia of the acute mono-myeloid type (M4) with a conspicuous skin infiltrate. The child was later referred to another hospital for treatment where she soon died.

Comments

Congenital leukemia with skin lesions was first described by Stransky in the year 1925.[2] Since then, several other cases have been reported.[2-6] Most of the cases of congenital leukemia have acute myeloid leukemia (AML). This is in sharp contrast with the occurrence of the acute lymphatic leukemia (ALL) in the first year of life. However, chronic myeloid leukemia and ALL are also known to present as congenital leukemia in the first week of life.[1] Eys and Flexner[7] proposed the following criteria for the diagnosis of CLC : (1) Manifestation at birth (in the first week of life), (2) Hepato-splenomegaly and lymphadenopathy, (3) Cutaneous involvement, (4) Elevated total leucocyte count with immature leucocytes in the peripheral blood, (5) Leukemic infiltration of the bone marrow, (6) Absence of erythroblastosis foetalis, syphilis and sepsis, and (7) Consistent autopsy findings.

Our case satisfied all these criteria except the last one. It is also important to differentiate a leukemoid reaction of unknown cause from the true AML. This can be done by repeated leucocyte counts and by examining the needle biopsies of the liver and spleen. Regression of leucocyte count to normal values (without antineoplastic treatment) and the absence of visceral infiltrates point to a leukemoid reaction.

The exact aetio-pathogenesis of CLC is not known. About 10% of CLC patients are reportedly associated with mongolism.[6] Transient congenital leukemia in seven infants with mongolism was reported by Engel et al.[8] In most of their cases, a complete and permanent remission was noted. The duration for which the leukemic cells persisted after birth in such cases are not recorded. It is interesting to note that neonates, whose mothers had suffered from leukemia during pregnancy, apparently do not manifest congenital leukemia.[5] Spontaneous and chemotherapy-induced remissions are known in CLC.[1]

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