

CONGENITAL LEUKAEMIA PRESENTING WITH SKIN NODULES

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A 2-month-old infant presented with the skin nodules of 1½ months duration. Routine investigation and bone marrow examination showed the presence of myeloblastic leukaemia-M₄ type, FAB classification.

Key Words : Congenital leukaemia, Myeloblastic, Acute myelogenous leukaemia (AML)

Case Report

A 2-month-old infant, born to non-consanguineous parents was brought to the hospital with the complaint of nodular skin lesions all over the body of 1½ months duration. The lesions had been increasing in size and number gradually. But for constant mild fever there was no other systemic complaint. No other relevant history was positive. On examination there were multiple, well-defined, round to oval, firm, mobile, non-tender, warm and erythematous nodules distributed over the scalp, face, neck, trunk and the limbs. Surfaces were smooth, shiny and infiltrated. Skin over the nodules could not be pinched. Darier's sign was negative. Differential diagnosis considered were

- Multiple mastocytomas,
- Eruptive histiocytomas,
- Juvenile fibromatosis and
- Congenital leukaemia.

The patient was admitted for investigations. However he expired the next day and the attendants refused post-mortem examination.

Investigations

Haemogram. WBC 67.93X10³/cu mm, RBC 2.94X10⁶/cu mm, HB 8.4 g/dl, HCT

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Fig . 1. Nodular lesions over the trunk.

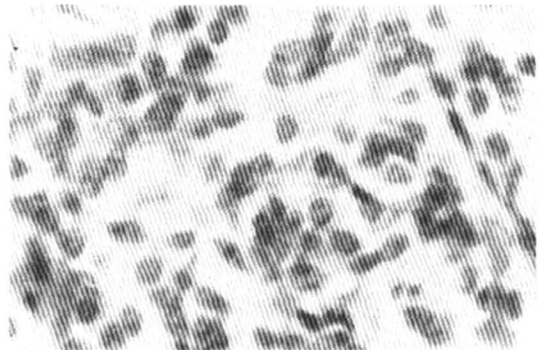


Fig. 2. Skin micrograph H & E 1000X. Note the features of AML-large cells, large nuclei and moderate cytoplasm.

26.4%, MCV 89.4 fl, MCH 28.4 pg. MCHC 31.6 g/dl, platelets 163X10³ and Reticulocytes 1.0%.

Differential count. Neutrophils 05, Lymphocytes 20, Monocytes 03, Eosinophils 02 and Blasts 70.

Histopathological examination of H & E section of skin biopsy specimen showed sheets of leukaemic blast cells in the dermis.

Bone marrow examination revealed sheets of large cells with large nuclei and moderate cytoplasm. 1-2 nucleoli were seen. Cytoplasm showed coarse granules. Some nuclei showed indentations and folds. Features suggestive of Leukaemia M₄ type, FAB classification.

Discussion

Congenital leukaemia is a well known condition. Acute myelogenous leukaemia accounts for 10-20% of childhood leukaemia. There are several conditions associated with increased incidence of acute myelogenous leukaemia (Table I).

Table I. Conditions associated with increased incidence of AML

Down's syndrome
Fanconi's syndrome
Bloom's syndrome
Kostmann's syndrome
Diamond - Blackfan anaemia
Drugs (alkylating agents, benzene)
Epidodophyllotoxins
Ionizing radiation
Myelodysplastic syndromes
Myeloproliferative syndromes

Increased incidence of M₄ and M₅ types of AML has been associated with 11q 23 deletions and translocations.¹ The most common symptoms at presentation of AML are anaemia, granulocytopenia, thrombocytopenia, fever, pallor, anorexia, weakness and cutaneous or mucosal haemorrhages.¹ Usually haemoglobin level between less than 3 to normal, low platelet count and absolute neutrophil count less than 1,000 are the findings.¹ Hepatomegaly and/or splenomegaly and/or lymphadenopathy can occur in more than 50% of the children with AML.²

Chloromas are solid collections of myeloblasts which can occur anywhere but are most common in the epidural and retrobulbar areas or in the skin, where they are termed leukaemia cutis.¹ Chloromas are most common in M₄ and M₅ types of AML.¹ Leukaemia cutis lesions are colourless or slightly purple ("Blue berry muffin" lesions).³ Occasionally chloromas are the only sign of disease and acute leukaemia almost always follows.⁴ Age less than 2 years, high WBC counts and M₄ & M₅ subtypes have been associated with higher incidence of central nervous system leukaemia.⁵ Most of the findings in our case suggest that the patient had congenital leukaemia M₄ type with skin lesions. However, as we lost the patient very early and the patient's attendants refused autopsy, we could not do extensive investigations/examination.

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