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Image Article

Acute Leukemic form of Blastic Plasmacytoid Dendritic Cell Neoplasm in Children

Stefano Carugo*

Department of Clinical Science and Oncology, University of Milan, Milano MI, Italy

ABOUT THE STUDY

A previously healthy 2-year-old girl had a fever and cough for 2 days. With 76% blasts, peripheral blood showed marked leukocytosis (white blood cells, 75000/mL) (Figure 1).

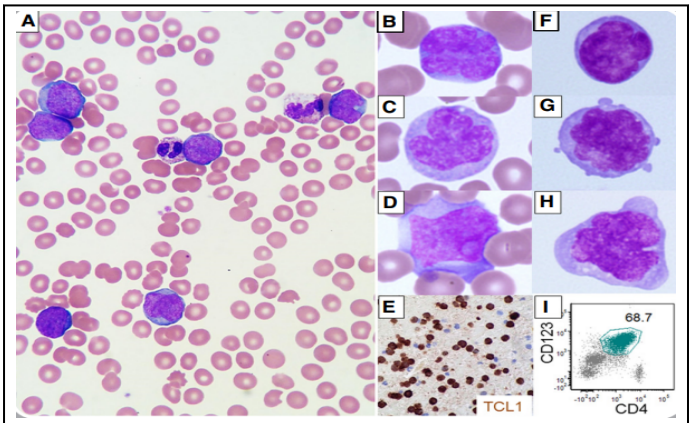

Figure 1: Blastic plasmacytoid dendritic cell neoplasm in Children.

Blastic plasmacytoid dendritic cell neoplasm in children

Panels A-D: The blasts varied in size from medium to large, with agranular paleblue cytoplasm. The nuclei were irregular, folded or formed like flowers. A prominent nucleolus was seen in some of the specimens. Blasts were found in 87% of the cerebrospinal fluid [1].

Panel E: 47, X, der(X)t(X;8)(q24;q24.1), add(5)(p13), -6, del(6)(q13q27), +8, der(8), t(X;8), add(8) (q24.1) × 2, del(9)(q12q31), del(9)(q13q22), der(10;13)(q10;q10), add(12)(p13), -14, +3mar (20), del(9)(q12q31), del(9)(q13q22), der(10;13)(q10;q10), add(12)(p13),-14 MYC rearrangement t(X;8) (q24;q24.1) is confirmed by Fluorescence *In Situ* Hybridization (FISH).

FISH (Fluorescence *In Situ* Hybridization) showed a single deletion of the *ETV6* gene but no MLL gene rearrangement, *BCR/ABL1* translocation, or *ETV6/RUNX1* translocation. The patient was diagnosed with blastic plasmacytoid dendritic cell neoplasm and was in complete remission 4 weeks after starting high-risk acute lymphoblastic leukaemia therapy, according to a bone marrow biopsy. The recognized distinguish this case: young age, negative CD^{56}, acute leukaemia without skin lesions, and central nervous system involvement [2].

Panels F-H: The lactate dehydrogenase level in the patient's blood was 3759 IU/L. The patient's lymphadenopathy was significant, but she had no skin lesions or hepatosplenomegaly. The blasts were positive for CD^{4}, CD^{7}, CD^{36}, CD^{38}, dim CD^{45}, CD^{123}, and HLA-DR by flow cytometry, but negative for the tested lineage markers CD^{2}, CD^{3} (surface and cytoplasmic), CD^{5}, CD^{14}, CD^{19}, CD^{20}, CD^{33}, CD^{34}, CD^{41}, CD^{56}, CD^{64}, CD^{117}, TdT, and myeloperoxidase by flow cytometry [3].

Panel I: The blasts tested positive for T-cell leukaemia 1 by immunohistochemistry.

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Correspondence to: Stefano Carugo, Department of Clinical Science and Oncology, University of Milan, Milano MI, Italy, E-mail: curugoitl12100@gmail.com

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