

P2. Dendritic cells acute leukemia – a rare disease with cutaneous onset

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Background. Dendritic cell leukemia (DCL) is a rare subtype of acute leukemia and is characterized by clonal proliferation of plasmacytoid dendritic cell precursors. Diagnosis by immunophenotyping allowed identification of this form of acute leukemia that was extracted from undifferentiated acute leukemia group. It is framed in the WHO classification of tumors of hematopoietic and lymphoid tissue editions in 2001 and 2008 as blast plasmacytoid dendritic cell neoplasm (BPDCN).

This disease has a characteristic immunophenotype with expression of following markers CD4, CD56, CD123, CD43. The skin lesions identified CLA expression. Also described is the expression of TCL1 (T-cell leukemia 1).

BPDCN is manifested by aggressive behavior that responds well to initial chemotherapy, but relapse is the rule and a poor prognosis. Clinical manifestations include cytopenia, particularly thrombocytopenia. In the literature there are few data about this subtype of leukemia, which makes it problematic to define the biological characteristics and clinical therefore appropriate therapeutic approach.

Material and methods. Results. We present a patient 84 years male who has been presented at diagnosis with maculopapular lesions posterior and anterior thorax and abdomen and splenomegaly.

Examination of bone marrow cellularity describes about 44% rich medium cells, the nucleus incised, fine chromatin, rare nucleoli, reduced quantity of basophilic agranular cytoplasm. Immuno-phenotypic analysis performed with FACS Calibur cytometer describes blasts population with CD45 low, SSC medium, with absence of stem cell markers CD34 and intracytoplasmic lineage markers, cCD3- cCD79a- cMPO- TdT-, myeloid markers, CD33- CD117- CD64-, lymphoid markers CD7- CD3- CD19- CD10- CD8-, except CD2+ CD4+/- CD56-/+ and coexpression of CD123+, CD36+ CD38+. Immunophenotypic markers were identified as meaningful for diagnosis of dendritic cell leukemia, according to the diagnostic score.

The diagnosis was supplemented by biopsy of skin lesions, which described rich cellular infiltrates perivascular and perianexial, consisting predominantly of lymphocytes in the dermis, with extension into the superficial portion of the hypodermis, concluding to the dermal inflammatory reactive nature in the context of a cutaneous pseudolymphoma.

During the outcome, the patient was followed clinically and by immunophenotyping, with favorable response to chemotherapy cures type CVP (cyclophosphamide, vincristine, prednisone), with the disappearance of splenomegaly and significant improvement of skin lesions while reducing component of dendritic cells from peripheral blood.

Conclusion. Dendritic cell leukemia is a rare group of acute leukemias and association with cutaneous lesions is suggestive of the diagnosis. Immunophenotyping is the main diagnostic tool that allows the diagnosis of this rare form of leukemia.