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Blastic Plasmacytoid Dendritic Cell Neoplasm with leukemic component

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A 72-year-old man was admitted to our hospital with a reddish skin tumor, which had appeared three weeks ago in his arm. The histopathological examination of a skin biopsy specimen was diagnosed as Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN). (Classification WHO2008) (figure 0). Physical exploration presented erythematous lesions on plaques, slightly indurated on the back and arms.

Three months later, the patient came to the emergency department complaining about dizziness and not feeling well. Physical exploration was anodyne. Complete blood count at this time showed a hemoglobin level of 10.2 g/dL, white blood cells 13.8 × 109 /L and platelets 51 × 109 /L. Peripheral blood smear demonstrated 97% of mediumsized blastoid cells with grey-blue cytoplasm, occasional cytoplasmic vacuoles, nucleoli, and some nuclear folds. (figures A, B) Bone marrow was packed (98% of cellularity) with immature cells negatively for myeloperoxidase, alphanaphthyl butyrate esterase and naphthol-ASD chloroacetate esterase stainings and positive reaction for the periodic acidSchiff. (figure C) Flow cytometry analysis showed blast cells expressing CD4/CD56/CD7/CD33/HLADR/dimCD45 with absence of CD34.

The diagnosis was BPDCN with involvement of peripheral blood. BPDCN is an aggressive hematologic malignancy originated from the precursor of the plasmacytoid dendritic cell. It has high frequency of cutaneous and bone marrow involvement and leukemic dissemination.

This neoplasm is a heterogeneous group of lymphoproliferative disorders, with different clinical, morphologic and immunophenotypic features.

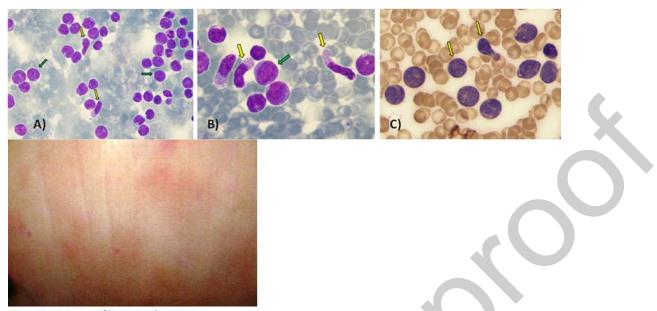


Figure-0: cutaneous infiltration at diagnosis