SKIN CANCER (OTHER THAN MELANOMA)

BLASTIC PLASMOCYTOID DENDRITIC CELLS NEOPLASM: CLINICO-PATHOLOGIC AND MOLECULAR DATA FROM A SINGLE MEDICAL CENTER.

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Introduction: Blastic plasmocytoid dendritic cells neoplasm (BPDCN) is a rare aggressive haematologic disease characterized by skin lesions (plaques and tumors) and simultaneous or subsequent involvement of peripheral blood, bone marrow and lymph nodes. BPDCN usually involves adults with a poor prognosis (median survival 23 months). Considering the rarity, no therapeutic guidelines are available but acute leukemia-type regime chemotherapy and stem cell transplantation are the mainstay of treatment.

Objective: The aim of this study is to collect clinico-pathologic and molecular data about a rare neoplasm from a single medical center.

Materials and Methods: We collected 32 cases of BPDCN and analyzed clinico-pathological data. Molecular analysis was performed using array-CGH on frozen DNA samples, from skin biopsy performed at the time of diagnosis.

Results: Our cohort includes 24 males and 8 females with a median age of 61 years (range 9-83). At onset, patients had cutaneous involvement in all cases with diffuse nodules and plaques in 25 of them and localized lesions in 7 of them. Seventeen patients also had bone marrow and/or peripheral blood involvement at the time of diagnosis. Median overall-survival (OS) was 24 months: 19 patients died from the disease, 2 died from the therapy and another death was not disease-related. All patients had typical peripheral dendritic cell phenotype. CD4 and/or CD56 were lost in 3 patients and aberrant expression of CD2 and/or CD7 was seen in 9 patients. A-CGH analysis revealed significant losses on chromosome 9 (68%), 13 (57%), 12 (12%), 7 (21%).

Conclusions: BPDCN is confirmed as an aggressive hematologic disease. Our study

confirmed histological, phenotypical and molecular data reported in our previous study. In particular, molecular analysis revealed several genomic losses involving multiple cell cycle checkpoints. The most frequent event was the deletion of 9p21.3, homozygously lost in patients with worse survival compared to longer survivors.