SKIN CANCER (OTHER THAN MELANOMA)

A RARE CASE OF BLASTIC PLASMACYTOID DENDRITIC CELL NEOPLASM IN A 43-YEAR OLD FILIPINO MALE: A CASE REPORT

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Background: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and highly aggressive hematodermic malignancy, with only approximately 100 cases reported worldwide. It usually occurs in elderly individuals with male predominance, and manifests with skin lesions followed by involvement of the lymph nodes, bone marrow, and peripheral blood. There is also a high risk for leukemic dissemination, hence, a poor prognosis.

Observation: A case of a 43-year old male is presented, with a 5-month history of solitary, well-circumscribed, non-movable, non-tender, 5.0 x 5.0 cm brown mass with overlying brownish-red papules, and surrounding erythematous plaque with follicular accentuation on the right upper back; and multiple erythematous patches and plaques on face, trunk, and upper extremities, with accompanying occasional easy fatigability, cervical lymphadenopathies, and pancytopenia. Histopathologic examination showed grenz zone with overlying flat epidermis, and nodular diffuse dermal infiltrates of atypical lymphocytes with hyperchromatic nuclei and prominent chromatin. Atypical cells extend around blood vessels and arrector pili muscle with atypical mitotic figures. Immunohistochemical profile showed positivity for CD56, CD4, Bcl-2, Ki-67, and S100; and negativity for CD3, CD20, CK20, CD45RO, CD99, CD10, and CD138. All of which were consistent with BPDCN – the first ever reported case in our institution.

Key message: A diagnosis of BPDCN is typically determined based on the histopathological and immunohistochemical examinations. However, despite the increasing number of cases reported in recent years, the diagnosis and treatment still remain a challenge due to the rarity of this diagnosis worldwide, as well as the clinical and phenotypical diversity observed among patients, and probable overlapping features with other hematologic malignancies. No standardized treatment regimen has been established as of this time; however, current evidence show that the preferred treatment regimen is that of intensive multi-agent chemotherapy with an acute lymphoblastic/myeloid leukemia (ALL/AML)-type protocol, followed by allogeneic hematopoietic stem cell transplantation (HSCT).