SKIN CANCER (OTHER THAN MELANOMA)

HYDROA VACCINIFORME-LIKE LYMPHOMA. A REVIEW OF SERIES OF 6 CASES FROM ARGENTINA IN CHILDREN

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Background: Hydroa vacciniforme-like lymphoma (HVLL) is an Epstein-Barr virus (EBV)-positive T/NK-cell lymphoproliferative disorder of childhood that occurs mainly in Central and South America and Asia. Clinically, it resembles hydroa vacciniforme (HV), presenting facial edema, papulovesicular lesions, blisters, ulcers, and varioliform scars that predominate on the face and extremities, but, unlike HV, it affects non sun-exposed areas. Fever, weight loss, lymphadenopathy, and hepatosplenomegaly are often observed. Histologically is characterized by perivascular and perianexal Epstein-Barr early RNA+ atypical lymphocytic infiltrate in the dermis and keratinocyte necrosis. Angiotropism, angionecrosis and subcutaneous involvement are occasionally present. The mayority of HVLL has a CD8+ T cell immunophenotype. CD56+ are rarely expressed. It may present clonal rearrangement of the T cell receptor gene.

Observation: We present 6 patients between 11 and 15 years old, with diagnosis of HVLLD. The mean age at diagnosis was 12.6 years-old, with predominance in males (ratio 5: 1). Clinically, all patients presented vesicles and generalized ulcers accompanied by facial edema, photosensitivity and hypersensitivity to mosquito bites. Five patients presented extracutaneous involvement: hepatosplenomegaly (3), lymphadenopathies (3), intestinal (1) and bone marrow (2) and hemophagocytic syndrome (1). All patients showed characteristic histopathological features. Regarding the immunohistochemical studies, all were CD3 +, 4 CD8 +, 1 CD4 + and 1 CD56+, 5 were EBER + and all had positive IgG EBV serology. A 4 of 6 patient EBV RNA was performed, and in all it was positive. All patients were treated with chemotherapy and one child also required radiotherapy. Currently, two patients died, one lost the follow-up, one patient is currently free of disease, one continuous follow-up in adult hospital and one remain on treatment

Key message: HVLL is an EBV-associated lymphoproliferative disorder of T/ NK-cell

phenotype with a broad clinical spectrum, usually prolonged clinical course, and risk for progression to systemic disease.