SKIN CANCER (OTHER THAN MELANOMA)

# INTESTINAL INVOLVEMENT IN A PATIENT WITH ADULT-ONSET HYDROA VACCINIFORME-LIKE LYMPHOMA

*K Férez-blando [1] - A Hernández-salazar [1] - G Agreda-vázquez [2] - Y Charli-joseph [3]*

*Instituto Nacional De Ciencias Médicas Y Nutrición Salvador Zubirán, Dermatology, Mexico City, Mexico [1] - Instituto Nacional De Ciencias Médicas Y Nutrición Salvador Zubirán, Hematology, Mexico City, Mexico [2] - Instituto Nacional De Ciencias Médicas Y Nutrición Salvador Zubirán, Cutaneous Hematopathology Clinic, Mexico City, Mexico [3]*

Background: A 34-year-old Mexican-mestizo man presented with a complex medical history beginning with a diagnosis of Crohn´s disease 5-years prior; based on the presence of intraepithelial lymphocytosis in intestinal biopsies, fever, diarrhea and weight-loss. In the following year, he developed numerous necrotic papules and hypochromic scars which slowly generalized to most of his skin, intermittent fevers, peripheral lymphoadenopathy and splenomegaly; leading to a skin biopsy-confirmed diagnosis of Hydroa vacciniforme-like lymphoma (HVLL). For a one-year period he received interferon α-2b with temporary improvement and subsequent progression. Upon his referral to our institution he was started on methotrexate, folic acid, prednisone and topical clobetasol with adequate tolerance, significant symptomatic improvement and resolution of most skin lesions. Four months later however, he developed abdominal pain, hypotension, tachycardia and fever. Two intestinal perforations were identified and infiltration of the intestine by EBV+ neoplastic T/NK-lymphocytes documented. He underwent ileum resection plus ileostomy and has recently started chemotherapy with l-asparaginase/metothrexate/dexametasone. He’s been admitted several times due to high-output ileostomy and acute kidney injury.

Observation: (HVLL), the aggressive end in the spectrum of hydroa-vacciniforme-like lymphoproliferative disorders (HVL-LPD), is a rare systemic NK/T-cell neoplasm, occurring mostly in Latin-American and Asian children with chronic active Epstein-Barr Virus (EBV) infection (CAEBV). It is easily misdiagnosed and has a 36% two-year survival. Whilst bowel perforation has been reported in 9% of primary gastrointestinal (GI) lymphomas it´s occurrence in lymphomas arising elsewhere is exceedingly rare. Regarding GI involvement in EBV+ T/NK-cell LPDs, a single case has been reported in a 14-year-old Chinese boy, with a fatal outcome and interestingly also misdiagnosed initially as Crohn´s disease.

Key message: The case reported herein highlight’s the salient features of HVLL and it´s rare potential for life threatening GI involvement.