DERMATOPATHOLOGY

ADULT-ONSET HYDROA VACCINIFORME-LIKE LYMPHOPROLIFERATIVE DISORDER

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Background: Hydroa vacciniforme-like lymphoproliferative disorder (HV-LPD) is a progressive cutaneous lymphoproliferative disease caused by chronic active Epstein-Barr virus (EBV) infection, including the spectrum from classic hydroa vacciniforme (HV) to severe HV and HV-like lymphoma (HVLL).

Observation: A 21-year-old female presented with recurrent necrotic papulovesicular lesions distributed on her face, chest and extremities for one year. The itchy papulovesicles appeared on the sun-exposed areas initially and gradually spread to sun-protected areas. No systemic symptoms such as fever, fatigue and weight loss were noted. Physical examination revealed multiple red-to-violaceous erythematous papules with central necrosis and hemorrhagic crust distributed on the face, chest, the upper arms, and pretrial areas. Results of complete blood count and comprehensive metabolic panel were unremarkable. Ultrasound revealed cervical and inguinal lymphadenopathy but no hepatosplenomegaly. Skin biopsy demonstrated a wedge-shaped lymphocytic infiltration with epidermal necrosis. Immunohistochemical staining showed the lymphocytes expressed CD3, CD8, CD30, and T-cell intracytoplasmic antigen 1 (TIA-1) with Ki-67 index being 50-60%. A diagnosis of HV-LPD was confirmed serologically with high levels of blood EBA DNA (over 4x107 copies/ml) in combination with Epstein-Barr encoding region (EBER) positivity detected by EBER in situ hybridization. TCR-γ and TCR-β gene monoclonal rearrangement were detected by polymerase chain reaction. The patient was treated with oral ganciclovir 250mg twice daily for 8 months with no significant decrease of blood EBV DNA level and frequent relapses after sun exposure. Interferon-α1b (30μg twice weekly, subcutaneous injection) was then added into the treatment regimen, leading to complete resolution and remarkable decrease of EBV DNA level to 1.76x104 copies/ml after 3-month therapy. Due to the possibility of HV-LPD for progression to systemic lymphoma, the patient was required to perform close follow-up with dermatology and hematology department.

Key message: Conservative therapy such as immunomodulatory treatment could be beneficial for HV-LPD despite the presence of T-cell monoclonality.