MEDICAL THERAPIES AND PHARMACOLOGY

CASE SERIES: THE TREATMENT OF HYPEREOSINOPHILIC DISORDER-LYMPHOCYTIC VARIANT WITH ROMIDEPSIN

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Background: Hypereosinophilic syndrome (HES) is defined as increased eosinophils in tissue or peripheral blood with an absolute eosinophil count >1500 cells/microL for at least 6 months without identifiable cause. In patients with the T cell lymphocytic variant of HES (LHES), clinical presentation often involves skin and soft tissue.

Observation: We present 2 cases of lymphocytic variant HES (LHES). Patient A is a 30-year-old lady who presented initially with chronic eczema and then found to have peripheral eosinophilia (absolute neutrophil count of 5.67 x109/L) and biopsy-proven cutaneous hypereosinophilia. Anti-neutrophil cytoplasmic antibody (ANCA) and strongyloides antibody were negative. FIP1L1-PDGFRA fusion gene was negative in peripheral blood. Peripheral blood flow cytometry displayed clonal T lymphocytes by analysis of the TCR beta-gamma receptor with a loss of CD3 and CD7. Cardiac MRI was negative for cardiac involvement by HES.

Patient B is a 77-year-old gentleman who initially presented with erythroderma and found to have severe peripheral eosinophilia (absolute eosinophil counts of 4 x 109/L). FIP1L1-PDGFRA and BCR-ABL fusion genes and assay for JAK2 V617F mutation were negative in peripheral blood. IgE levels were markedly elevated (initially > 3000 IU/mL) and toxocara serology was positive. Patient completed a course of albendazole without relief of erythroderma or pruritus.

Key Message: Both patients had minimal response to steroids, but experienced improvement in skin after 1 cycle of romidepsin. Romidepsin is a histone deacetylase inhibitor approved for Peripheral T-cell Lymphoma and Cutaneous T-cell Lymphoma (CTCL). The mechanism of action results in a decreased cell growth of T-lymphocytes in the setting of increased apoptosis induced by DNA damage. The two patients we are reporting had dramatic response to romidepsin with almost complete resolution of their symptoms suggesting that the biology of LHES may share resemblance to CTCL.