HYPERIMMUNOGLOBULIN E SYNDROME AS A CAUSE OF SEVERE CHRONIC INFECTIOUS SKIN LESIONS: A CLINICAL CASE

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Abstract. Hyper-IgE syndrome (HIES, Job syndrome) is a congenital immune system disorder (primary immunodeficiency) that is characterized by a combination of extremely elevated levels of total immunoglobulin E, impaired cellular immune response, and a predisposition to severe recurrent infections and allergic diseases. This article presents a clinical case of primary combined immunodeficiency with late-onset hyperimmunoglobulin E syndrome, the course of which was accompanied predominantly by infectious skin lesions. Clinical presentation included chronic pyoderma, microbial eczema, the development of necrotic trophic ulcers of the upper and lower extremities, as well as the development of hypersensitive leukocytoclastic vasculitis. Immunological testing revealed a combined defect in T-, B-, and NK-cells, combined with persistent hyperimmunoglobulinemia E and polysensitization, which corresponded to the immunopathogenic profile of hyper-IgE syndrome. Intravenous immunotherapy with Normal Human Immunoglobulin (NHIg) has resulted in a significant clinical presentation improvement, a decrease in the activity of the pathological process in the skin and stabilization of the patient's general condition. The presented clinical case highlights the diagnostic challenges of HIES in case of late onset and underscores the need for comprehensive immunological and molecular-genetic testing to optimize treatment strategies, predict disease progression, and improve the patient's quality of life.

Key words: Hyper-IgE syndrome, HIES, Job syndrome, primary combined immunodeficiency, hyperimmunoglobulinemia E, chronic pyoderma, trophic ulcers, replacement immunotherapy.